RHD Action | United to End Rheumatic Heart Disease

RHEUMATIC HEART DISEASE CONTROL PROGRAMMES

Tools for Implementing Rheumatic Heart Disease Control Programmes

TIPs HANDBOOK

SECOND EDITION

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Medtronic FOUNDATION

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PREFACE TO THE SECOND EDITION OF TIPs

An estimated thirty-three million people live with rheumatic heart disease (RHD) globally. They live in low-resource settings and in vulnerable communities worldwide. For too many of these people, RHD causes symptoms in adolescents and early death in young adulthood. RHD extracts an additional toll on women during pregnancy and is a major contributor to maternal mortality in endemic countries. The impact of RHD on individuals, families and communities is tremendous and it is preventable.

Almost all of the clinical, scientific and programmatic knowledge needed to care for people living with RHD, and to prevent new cases of RHD already, exists. We know that RHD can be controlled. The challenge is to deliver comprehensive RHD programmes which build on this existing knowledge, amplify best practice and share our successes. This is not a disease amenable to a single intervention. RHD control requires sustained prioritisation and commitment across the health care spectrum – from sore throat infections in clinics to open heart surgery in regional centres. It exemplifies the intersectionality of contemporary disease control challenges.

This second edition of Tools for Implementing RHD Control Programmes (TIPs) provides a platform for RHD control programmes in low resource settings. Primarily it is written for people on the frontline of disease control decisions – programme managers and policy makers. Their efforts and commitments have underpinned many of the historical successes in RHD control. By offering an overview of programmatic issues TIPs makes it possible to begin renewed conversations about local strengths, priorities and pathways to tackle RHD.

Successfully controlling RHD also requires action which is outside the scope of TIPs. The social determinants of health must be addressed to improve the conditions in which people are born, grow, live, work and age. Clinical best practice must be codified into guidelines to improve medical management. The voices of people and communities living with RHD must be elevated to determine priorities and progress in disease control.

The second edition of TIPs has been published in May 2018, ahead of the 71st World Health Assembly which will consider a resolution on rheumatic fever and rheumatic heart disease. The proposed resolution promises a new era for global RHD control, emphasising the practical action and political prioritisation outlined in this edition of TIPs. We believe that this will be a new beginning in ending RHD around the world.

TIPs Second Edition Writing Committee

INTRODUCTION

Each year, more than 300,000 people die from rheumatic heart disease (RHD).

Almost exclusively, the people who die of RHD live in low- and middle-income countries or in vulnerable communities in high-income countries. Their deaths are preventable with medical knowledge and antibiotics which have existed for more than half a century. In high resource settings socioeconomic and medical determinants have functionally eradicated RHD. Yet preventing, diagnosing and treating RF and RHD remains a fitful struggle in low resource settings. Death and disability from RHD continues to extract an enormous social, economic and cultural toll on young adults and their communities. The burden is greatest in the most productive years of life for those who can least afford it. The absolute burden of disease, the social effect, economic cost and the abject inequality of RHD demand urgent global action. TIPS provides a resource for people and places contemplating an RHD control programme. The collation of decades of implementation experience from around the world provides a solid foundation for customised programme development. TIPs presents an overview of RF, RHD and opportunities for intervention, alongside a priority-based framework for programme delivery. The resource is intended to support the description, development and delivery of RHD control programmes.

Overview

Sore throat (pharyngitis) is a common childhood infection in most parts of the world. The majority of sore throats are short viral infections which resolve without complication. However, up to 30% of sore throats are caused by a bacterial infection. The most common cause of bacterial sore throats is Strep A infection. In susceptible young people Strep A infections of the throat can cause an abnormal immune reaction, rheumatic fever (RF). This abnormal immune response causes inflammation of the heart (carditis) and, with repeated Strep A infections, scarring of the heart valves. Damage to the heart valves is called RHD. Over time, the heart valves become too scarred to function, causing heart failure and increasing the risk of abnormal heart rhythms, heart valve infections and complications during pregnancy.

At least 33 million people live with RHD around the world and 319,000 people die annually of the disease.[2] Overwhelmingly these deaths are premature and occur in young adults. Most people dying of RHD are aged under 40 years.[3-5]

The vast majority of people with RHD live in developing countries.[2] Others live in high resource countries in Indigenous communities and other vulnerable populations. The socioeconomic distribution of RHD reflects its root cause in poverty, household crowding, inequality and inadequate access to medical care.

Even in very low resource settings the prevalence of RHD reflects a socioeconomic gradient; this is a disease which afflicts the poorest of the poor. Poverty amplifies the tremendous human, social and economic burden of RHD. Acquired in childhood or adolescence, RHD reduces school attendance and education outcomes.[6,7] People with symptomatic RHD are less able to work at the same time as their healthcare costs increase. In endemic settings, people living with RHD often bear the economic cost of accessing health services and medication. Heart surgery can reduce symptoms and prolong life with RHD but is unavailable in settings where the disease is most common and prohibitively expensive in many other places.[5,6] Women with RHD are at far greater risk of death during pregnancy and labour, contributing to the intergenerational transfer of poverty and causing complex social, cultural and relationship harm.[8,9] The profound inequality of RHD amplifies the social, economic, pragmatic and humanitarian rationale for disease control.

The burden of RHD is the number of people developing, living with, and dying from, the disease (incidence, prevalence, disability and mortality). The burden of RHD also refers to the impact of the disease on individuals, families, communities and governments. RHD control encompasses prevention, diagnosis and treatment of RHD to reduce the burden of the disease.

Disease control is challenging because it requires the community, health system and government to work together in a coordinated way. Coordination must be maintained for many years to influence the number of people developing RHD and reduce the number of people living with the disease. An RHD control programme is a structured plan to prevent, diagnose and treat Strep A infections, RF and RHD and to reduce the burden of the disease.

RHD control programmes have been implemented around the world for more than sixty years.[10] Most programmes have included a list of people living with RHD (an RHD register) in order to provide secondary prophylaxis with antibiotics to people at risk of recurrences of RF. Others have focused on primary prevention by treating sore throats with antibiotics to reduce the risk of RF and subsequent RHD. Delivery of these services often requires health system interventions, including health worker training, government engagement, disease notification systems and securing antibiotic supplies. RHD control programmes may also incorporate medical management of people with symptomatic RHD and facilitate access to cardiac surgery. Other roles commonly include research and epidemiology to understand the burden of disease and advocacy to address the underlying social determinants of disease.

TIPs collates the implementation experiences of these RHD control programmes from around the world to provide an overview of approaches to RHD control. The handbook is intended as a 'menu of options' for comprehensive disease control programmes, addressing considerations for each component.[11] The relevance of each component will be determined by local needs, priorities and experience.

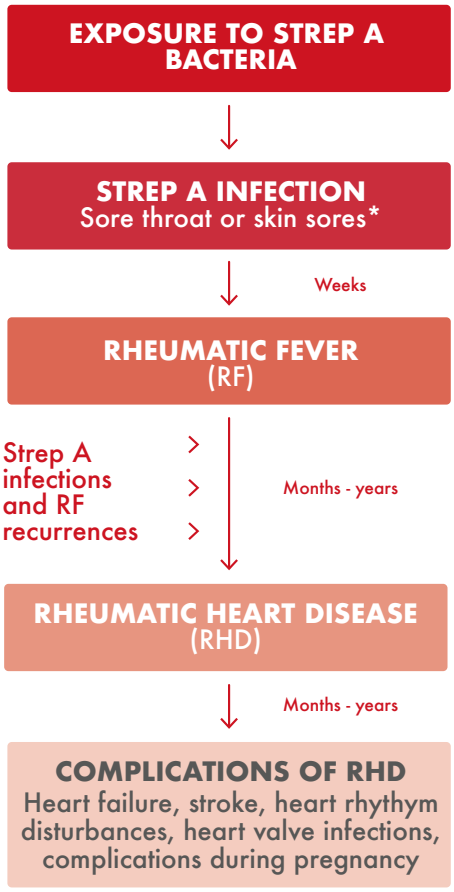


Figure 1: Causal pathway of RHD

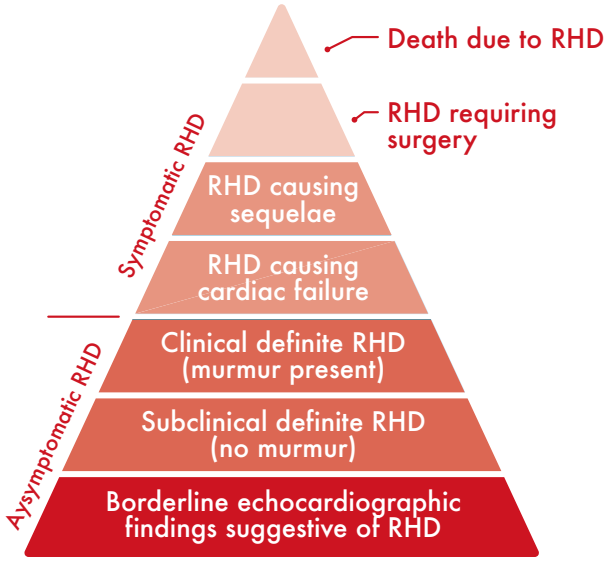


Figure 2: Population model of RHD progression

What are RF and RHD?

Most sore throat infections in children and young people are caused by viral infections. A variable minority (0-30%) of sore throat infections are caused by Strep A bacteria (sometimes known as Group A Strep, GAS, and Streptococcus pyogenes).[12] Strep A also causes skin and soft tissue infections. Some people have an abnormal immune reaction to Strep A infection. This abnormal immune reaction means that, as well as attacking the Strep A infection, other parts of the body are also attacked – causing RF.

RF causes joint pains, fever, skin changes and sometimes abnormal movements (chorea). In most cases the heart also becomes inflammed during RF (carditis).[13] However, when other symptoms of RF resolve, changes to the heart valves persist. Repeated episodes of Strep A infection and RF cause progressive heart valve damage. This persistent valve scarring is called RHD.

The risk of RF following untreated Strep A pharyngitis is between 0.3 and 3%.[14-16] For individuals with a history of previous RF the risk rises to 50%.[17] The number of recurrences of RF are an important determinant of disease progression.[18]

The classical pathway of individual progression from Strep A infection to RF and RHD is illustrated in Figure 1. This diagram is a good, simple way to understand the disease. Advances in echocardiography have revealed that the reality is probably a little more complex – a latent phase of subclinical RHD precedes clinical signs and symptoms.[19,20] A diagram of disease progression at a population level appears in Figure 2.

Only some people are susceptible to RF and RHD. A triad of environmental, genetic and bacterial factors appear to be important in the development of clinically significant disease.[21] These mechanisms are the subject of ongoing biomedical research and are not addressed in this handbook.

What is a comprehensive RHD control programme?

There are many opportunities to intervene on the pathway from Strep A to RHD. Traditionally these have been divided into primordial, primary, secondary and tertiary interventions - illustrated in Figure 3.

Register-based programmes for RHD control have been recommended by the World Health Organization (WHO) and World Heart Federation (WHF) for many years.[22,23] In reality, most programmes are more than a register – they include efforts to treat sore throats, educate communities, engage people living with RHD, support healthcare workers and treat the complications of advanced disease.[24] These programmes are sometimes called 'comprehensive' because they include primary, secondary and tertiary prevention. Although different components of comprehensive programmes may develop at different stages it is increasingly clear that primary and secondary prevention work synergistically to reduce the burden of RF and RHD.[25-27] In addition to primary and secondary prevention, ethical and humanitarian considerations mandate inclusion of medical and surgical services for people already living with RHD.

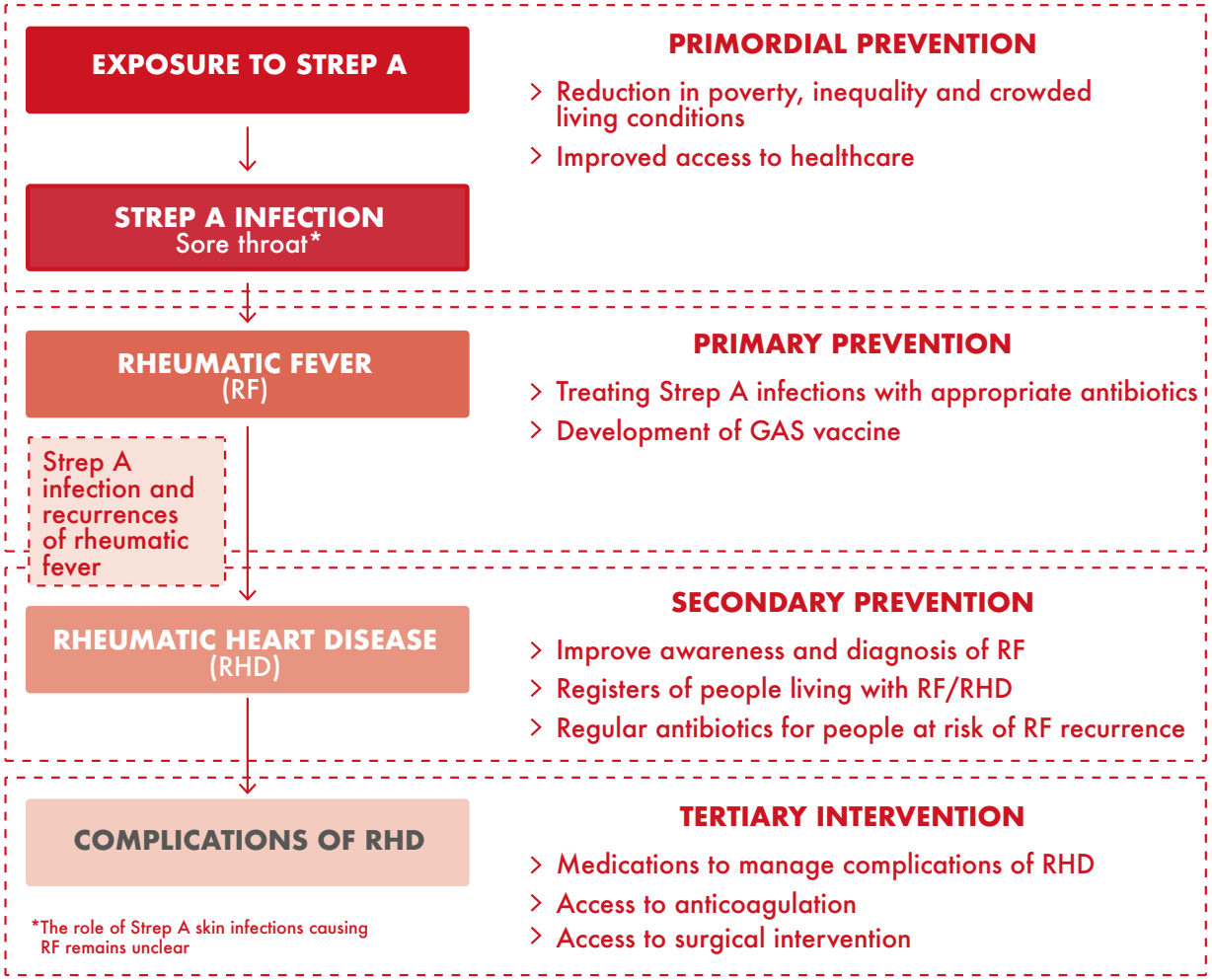


Figure 3: Opportunities for intervention in RF and RHD

Comprehensive RHD control programmes necessarily include a broad range of priorities, decisions and tasks. A conceptual framework is useful for visualising some of these issues – a proposed conceptual framework for comprehensive RHD control programmes is illustrated in Figure 4.[11] This implementation focused framework provides a structure for the following TIPs chapters. The framework was formed by reviewing WHO recommendations and other programme implementation experiences to identify common critical element for RHD control.[11]

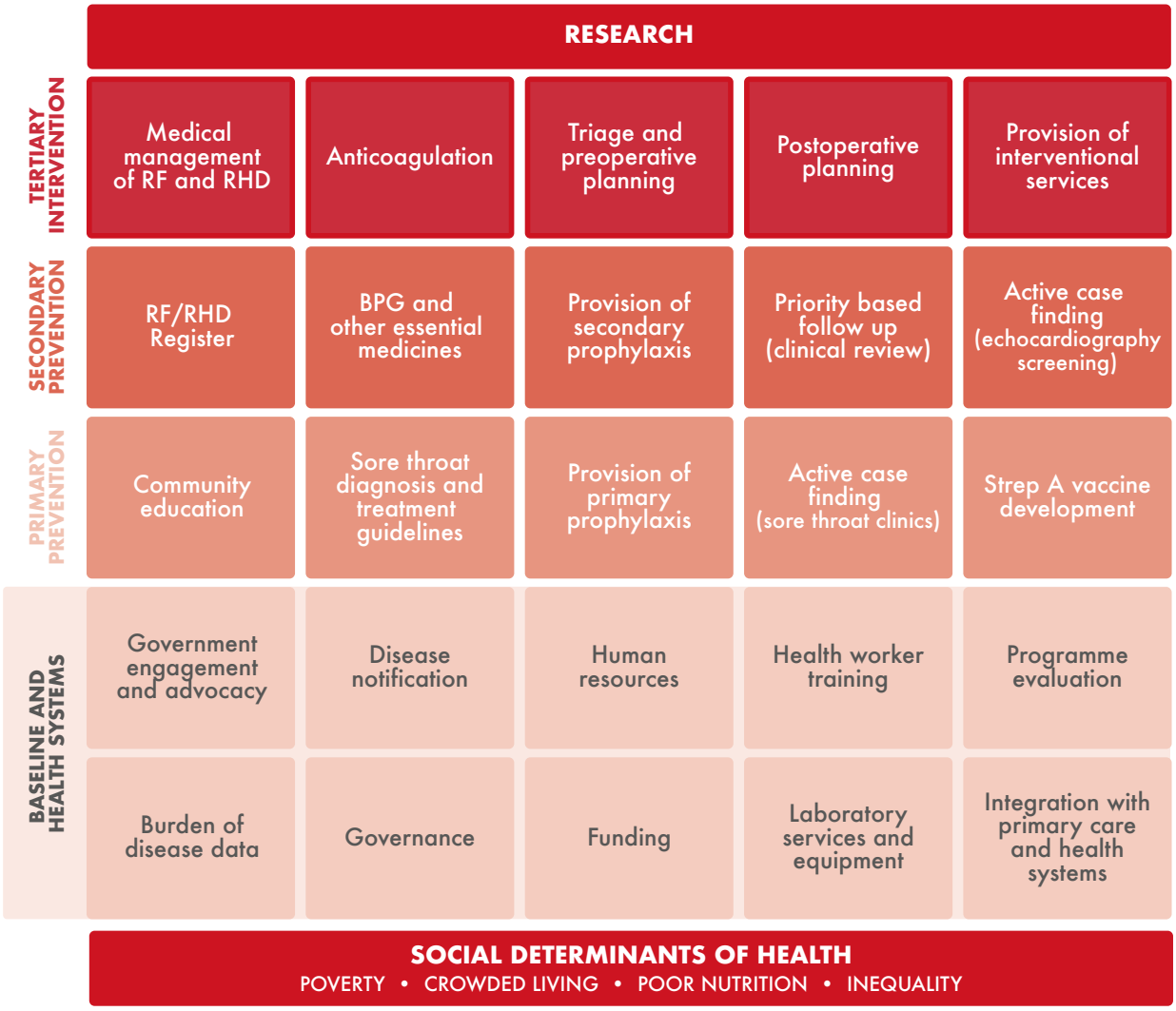


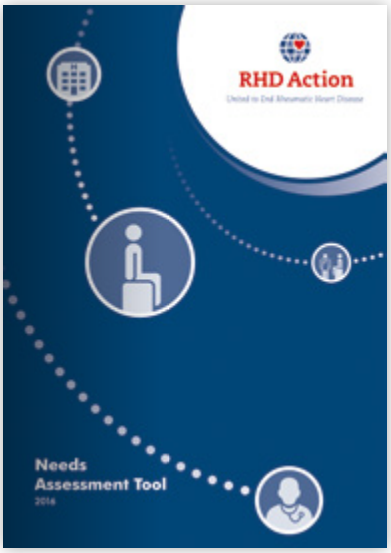
Figure 4: A conceptual framework for comprehensive RHD control programmes.[11] Components are arranged in approximate order of priority, working from left to right, bottom to top, in each row

The approach illustrated in Figure 4 offers a way of describing, designing, implementing, and evaluating comprehensive RF/RHD control programmes. The components are arranged in approximate order of priority, working from left to right, bottom to top, in each row.[11] This conceptual framework emphasises the need to tackle core components (i.e. antibiotic supply) before more complex interventions (i.e. echocardiographic screening and cardiac surgery). Therefore, initial priorities for a new programme are suggested in the left-hand column – burden of disease data, government engagement, community education, development of an RF/RHD register and medical management of people already living with RHD.

In practice, development of comprehensive RHD control programmes tend to follow a rough trajectory. Champions – often clinical staff – in one geographic location are aware of the burden of RHD – seeing first-hand the impact on families and communities. They gather stories and existing data in their attempt to engage other people on the issue. Typically, this reveals a lack of information about the prevalence of RHD in their setting and champions begin to undertake some kind of research to better understand the burden of RHD. In the process, a register is often developed to help count the number of people living with the disease and to manage patients receiving secondary prophylaxis. In developing settings with a high burden of disease, most people newly identified with RHD have advanced disease and are very unwell. As more people become aware of the poor outcomes of RHD, and learn that the disease is entirely preventable, there is increasing momentum for action. Register-based care begins to formalise and the register expands to include care in other geographic locations.

This organic trajectory has many determinants, often spurred on by passionate individuals, sporadic access to surgical services, visiting teams, technical support, political engagement and media coverage. The process can be derailed by fatigue, resource constraints, shortages of essential medicines and competing health and political priorities.

RHD Action has developed a Needs Assessment Tool (NAT) which can be used alongside TIPs to support the trajectory of developing comprehensive RHD control programmes in resource limited settings.[28] The NAT is a collection of over 30 data collection, programme development, and monitoring and evaluation template forms to assess baseline capacity and identify key areas for interventions. The model is based on understanding the needs of a candidate site to inform future programme development and/or expansion as outlined in Figure 5.



RHD Action Needs Assessment Tool

Figure 5: Stages of the RHD Action Needs Assessment Tool

Opportunities to use elements of the NAT are identified throughout this book. The full resource can also be downloaded from rhdaction.org.

Some elements of the NAT approach have been used in RHD Action Demonstration Partners in Uganda and Tanzania. A sample baseline systematic review of RHD in these countries was published in 2017.[29] The review explored 3 broad objectives:

- To quantify the burden of Strep A, RF and RHD in Tanzania and Uganda.
- To describe the patient and provider experience with Strep A, RF and RHD in these countries.
- To identify the types of stakeholders who currently are, or need to be, engaged when designing and implementing RHD programmes in these communities.

This review demonstrates the utility of taking stock of what is already known about RHD in one setting and using that information to inform programme development.

The conceptual framework of TIPs is not designed to be prescriptive and you certainly don't need to tackle everything. The details of designing and delivering RHD care will be unique in each setting. Your local needs are the most important consideration: community consultation, existing infrastructure, political and economic feasibility of programmes and human resources. The framework in Figure 4 is simply a tool to help structure your thoughts about what needs to be done and in approximately which order.

Using TIPs

TIPs is quite a long document and not designed to be read from beginning to end in one go. Different parts will be relevant to your programme at different times and for different people. The 'Things to consider' section at the beginning of each chapter summarises some of the main points – you may like to review these questions before deciding whether to spend more time on each chapter. 'Opportunities for integration are also highlighted', identifying scope for your programme to work with other diseases, programmes or departments to improve care delivery.

Who should use this handbook?

TIPs is written primarily for people implementing RHD control programmes, particularly programme managers and clinical advisors. However, we hope that the handbook will be a useful reference for everyone engaged in RHD control. You might be an interested doctor, a nurse, a teacher, a policy maker or someone living with RHD. You could be a group of people beginning to plan a control programme. You may want to evaluate an existing programme or participate in a visiting surgical team to a setting with a high burden of RHD. The text is designed to be relatively accessible to anyone interested in reducing the burden of RF and RHD in their community.

You do not need to have any special training to use the TIPs handbook. There is a plain text summary of some of the medical issues in control of RF and RHD on page 2. If there are things you think should be clarified or better explained in future editions we'd love to hear from you – contact details appear inside the cover.

Methods and limitations of TIPs

TIPs collates 60 years of programmatic experience delivering different components of comprehensive RHD control programmes. The first edition of TIPs was produced in 2014. This 2018 second edition has been updated to reflect new knowledge and best practice.

The core references were identified through a systematic literature review of EMBASE, BIOSIS and PubMed searches of English and French articles from 1952 to 2014 and then from 2014 to February 2018. Search terms included: "rheumatic" AND (heart disease OR fever NOT arthritis) AND (control OR prevention OR prophylaxis) AND (progra* OR strateg*)" plus focused searches for specific components of control programmes, including: regist*, community education, training, anticoagulation and disease notification and surveillance. Article titles and abstracts were reviewed to evaluate suitability for inclusion. Sentinel articles were selected for bibliographic review to identify additional references, personal communications or unpublished reports.

Unpublished or informal 'grey literature' was identified through research and programme collaborators of the writing committee. Additional Google searches for programme reports, evaluations and non-database indexed references were conducted. A snowball approach was used to identify other source documents accessible through direct contact with individuals and institutions.

Although review of the existing RHD programme delivery literature has been extensive, the TIPs handbook has a number of limitations:

- RF/RHD remains a disease of vulnerable populations, often living in resource limited settings where research and information sharing is limited. This inequality of evidence is changing slowly with increasing amounts of high quality information available from low resource settings.[30] However, many questions critical to the management and control of RF and RHD remain poorly understood. Some evidence still comes from historical studies in relatively high-income countries from the 1950s to 1970s. It is unclear whether these experiences can be directly extrapolated to currently endemic, low resource, regions.
- Some components of comprehensive disease control programmes have not been described or analysed in sufficient detail. For example, there are relatively few papers on integrating RHD into the broader health system or interfacing with surgical services. We have tried to share the experience of other relevant disease programmes where possible but this remains an outstanding research area.
- Literature review was limited to English and a small number of French language resources. Experience from non-English settings is likely to be under-represented. Similarly, search strategies were conducted largely online; this electronic dependence has produced a relative over-representation of references from high income settings with a burden of RHD in vulnerable populations (particularly Australia and New Zealand).
- Many of the areas addressed in TIPs are independent fields of research and implementation. For example, laboratory management, programme evaluation, recruitment and retention of health workers are all specialty domains in their own right. We have summarised key issues in these domains and provided additional references for more detailed information.

RHD CONTROL IS ACHIEVABLE

The task of reducing RHD can appear overwhelming. However, the achievements of landmark programmes demonstrate that significant progress is possible. The feasibility of disease control was highlighted in a 2017 editorial on RF and RHD in a special edition of the World Heart Federation journal, Global Heart.

> "Imagine a future scenario, a world in which all group A streptococcal pharyngitis is readily identified at the point of care, definitive acute treatment is prescribed, the effective medication is readily available and affordable in all health care settings, and the prescribed long-term secondary prophylaxis is widely acceptable and affordable from the patient's perspective. That future is already here, but as William Gibson so eloquently put it, "it's just not very evenly distributed."
>
> Mensah et al, RF and RHD Research, 2017.[31]

THE REPUBLIC OF COSTA RICA EXPERIENCE

Costa Rica is a Central American country with a 2015 population of 4.8 million people.[36] In the 1950s and 1960s RF and RHD were a major health burden –the attack rate of RF in 1950 was 120/100,000 people and a quarter of deaths in 5–14 year olds were attributable to RF/RHD.[35] In the 1970s efforts to strengthen access to primary care in the country included a scale up of primary prevention of RF. The requirement for a positive Strep A throat swab before treatment of pharyngitis was removed. Clinical guidelines were changed to make a single dose of injectable antibiotic first line therapy for suspected Strep A pharyngitis. An education campaign was developed for healthcare providers and an increased supply of antibiotics was provided to health clinics.[37] Following these changes, the incidence of RF fell and presentations to the National Children's hospital reduced from 94 new cases of RF in 1970 to 4 new cases in 1991 (see Figure 6).

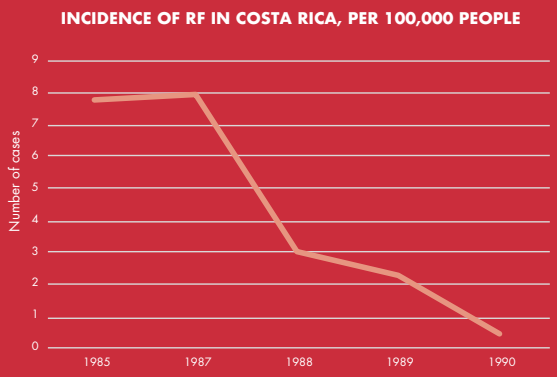


Figure 6: Incidence of RF Costa Rica 1985 – 1990[35]

THE PINAR DEL RIO CUBAN EXPERIENCE

Pinar de Rio is a province at the Western end of Cuba, an island nation in the Caribbean Sea which had a population of 721,800 in 1996.[32]

Baseline data on the burden of RF and RHD were collected in Pinar del Rio in the 1970s and 1980s. A comprehensive control programme began in 1986, including primary and secondary prevention of RF/RHD with the following components:[33]

- Educational activities and training workshops were organised at provincial, local and health facility level.
- Health education and dissemination of information. 'Thousands of pamphlets and hundreds of posters were distributed, and special programmes were broadcast on the public media to advertise the project.'
- Community involvement.
- Epidemiological surveillance.
- Permanent local and provincial RF/RHD registers were established at all hospitals, polio clinics and family physicians in the province.

By 1996 the incidence of RF had fallen from 18.6/100,000 to 2.5/100,000, an absolute risk reduction of 83%. Rates of RF recurrence also fell by 94%. This reduced burden of disease persisted until at least 2002, even when the control programme had formally concluded (see Figure 6).[33] In 2015 a new cost effectiveness analysis of the Pinar del Rio approach was conducted and the programme was found to be cost saving.[32] The effect of both primary and secondary intervention contributed synergistically to this outcome. The authors note that, 'the results of our analysis suggest that, even in low-resource settings, inaction on RHD is much more costly than action'.[32]

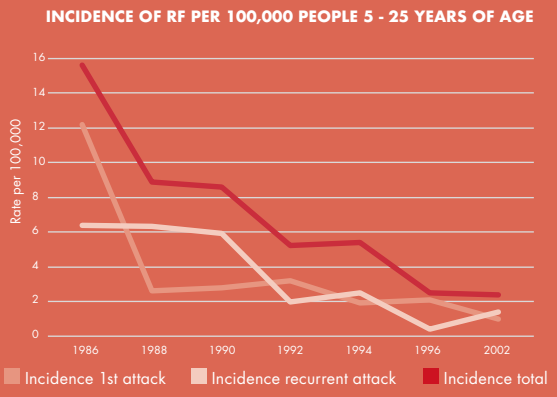


Figure 7: Incidence of RF, Pinar del Rio 1986 to 2002[32]

THE NEW ZEALAND EXPERIENCE

New Zealand is a highly developed country in the Pacific Ocean, with a population of 4.5 million people in 2015.[39] Indigenous Māori and Pacific Island communities within New Zealand have high rates of RF and RHD. In 2012, reducing the rate of RF became a Better Public Service (BPS) target – this political prioritisation mandated different government departments to work together to address RF. The target was to reduce the incidence of RF by 66% from a baseline of 4/100,000 in 2009/10 – 2011/12 to 1.4/100,000 by 2016.[40] The programme had three main strategies:

- Increase awareness of RF, what causes it and how to prevent it.
- Reduce household crowding and therefore reduce household transmission of strep A bacteria within households.
- Improve access to timely and effective treatment for strep throat infections in priority communities. This included both school-based and primary care sore throat management and primary care sore throat management.

By December 2016, the incidence of RF in New Zealand had fallen by 23%, illustrated in Figure 8.[38]

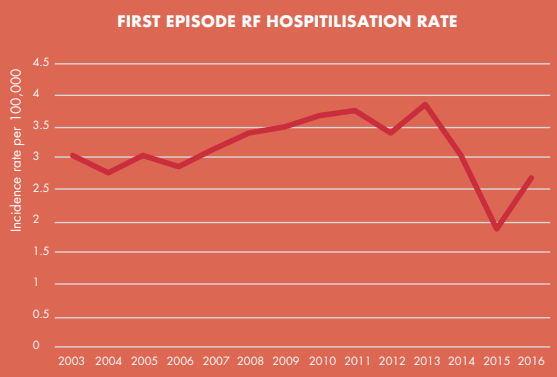


Figure 8: Incidence of RF admissions in New Zealand 2002 – 2016.[38]

THE MARTINIQUE AND GUADELOUPE FRENCH CARIBBEAN EXPERIENCE

In 1981 an RHD control programme was established in two French Caribbean islands, Martinique and Guadeloupe. The islands were middle income settings with relatively strong health systems and free access to healthcare and medication.[34] The programme had four key principles:

- Development of a register.
- Health worker and community education.
- Research.
- Treatment of skin infections.

A full time paediatrician dedicated to RF was employed in each island. By 1992 the incidence of RF had declined by 78% in Martinique and 74% in Guadeloupe (see Figure 9). The cost to the health systems of RF reduced by 86%.[34] The authors found that, 'Our major conclusion is that rapid decline in rheumatic fever incidence can be achieved with few staff at modest cost.'[34]

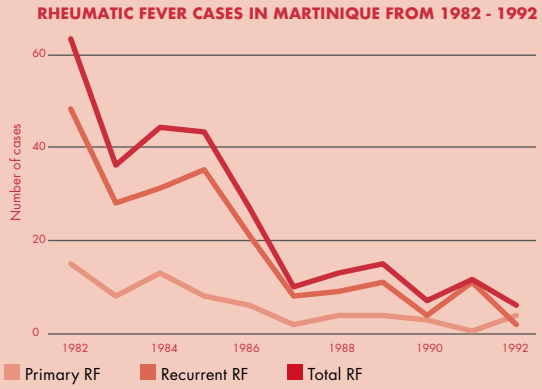


Figure 9: RF cases in Martinique from 1982 - 1992[34]

THE NORTHERN TERRITORY AUSTRALIA EXPERIENCE

The Northern Territory in highly-developed Australia is home to 220,000 people.

Twenty-five percent of the population identify as Indigenous Aboriginal or Torres Strait Islander people.[42] In 1997 the Top End of the Northern Territory (NT) region of Australia developed a register based programme for control of RF and RHD.[43] RF is a notifiable condition in the region and active surveillance is maintained through health professional education and a small number of dedicated programme staff.[44] All known cases of RF, recurrences and RHD are entered into a Territory-wide computer-based register.[45] In 2017 there were approximately 3000 people on the central register, the overwhelming majority of whom are Aboriginal Australians.[41] People on the register are managed according to comprehensive national clinical guidelines including regular 4 weekly antibiotic injections if indicated.[46] Public health nurses employed by the NT RHD Control Programme travel the Northern Territory and provide support to primary healthcare centres in the development and delivery of services. A large proportion of these primary care clinics are in remote locations. Nurses provide training and education to health staff, patients and their families. A programme review in 2013 provided evidence of programme success: the recurrence rate has fallen by 9% per year since the programme began in 1997.[47] Adherence with secondary prophylaxis injections has increased – in 2008 only 18% of patients received 80% of annual scheduled doses, in 2017 46% of people are receiving greater than 80% of injections (See Figure 10).[41]

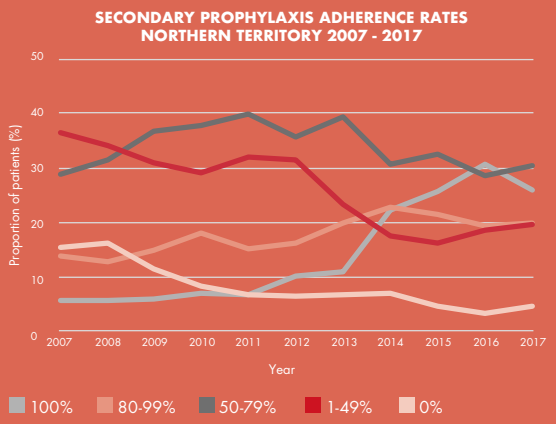


Figure 10: Adherence to secondary prophylaxis in the Northern Territory of Australia.[41]

THE MINAS GERAIS EXPERIENCE

Minas Gerais is a state in the upper-middle income country Brazil, home to 19.6 million people.[48] A comprehensive RHD control programme has been administered by the Reference Centre for Rheumatic Fever since 1988. The programme includes the following core components:[48]

- Establishment of the Rheumatic Fever Outpatients Clinic.
- Introduction of protocols for standardised approach to diagnosis and management.
- Medical care and orientation by multidisciplinary teams.
- Health promotion to people living with RHD and their families.
- Centralisation of appointment scheduling
- Clinical follow up twice yearly with free transport provided.
- Access to free medications including benzathine penicillin G every 21 days.
- Accommodation provided for parents of admitted inpatients.

Longitudinal records of patients from this programme were reviewed in 2015.[48] The study authors report that adherence with secondary prophylaxis improved, although the improvement in adherence was not quantified. Changes in clinical outcomes were assessed by comparing outcomes from the period 1977–1978 with the period 1988–2000. A statistically significant reduction in recurrences, severity of carditis, severity of valve disease, need for surgery and death was demonstrated. In this setting a comprehensive RHD control programme - with a focus on secondary prevention – appears to have substantially improved clinical outcomes for children with RHD. This appears to be reflected in declining admissions for RF and RHD throughout Brazil. However, a separate echo screening study in Minas Gerais suggests that the population prevalence of RHD remains very high at 42/1000 school students.[49]

BACKGROUND CLINICAL INFORMATION

This section provides a plain text summary of medical conditions relevant to RF and RHD. It is provided to assist people without medical training to become familiar with medical issues in RF and RHD control. Additional information for clinicians is provided in Chapter 21 – Medical management of RF and RHD.

Complications of RHD

Heart failure

The major cause of death and disability from RHD is heart failure, sometimes called congestive heart failure (CHF). Over time, scarred and damaged heart valves (usually the mitral valve) cause pressures to rise within the chambers of the heart and the heart to fail as a pump. Without a well-functioning heart, fluid builds up in the lungs and body, causing symptoms of breathlessness, swelling and fatigue. These symptoms tend to become worse over time without treatment.

Stroke

Stroke occurs when a part of the brain does not receive adequate blood supply. Strokes can be ischaemic (from a blocked blood vessel) or haemorrhagic (from a burst blood vessel). People with RHD are at risk of ischaemic stroke because of blood clots which can form in the heart and subsequently block blood flow to parts of the brain. Some people living with RHD need to take 'blood thinning' medication (anticoagulation) to reduce the risk of stroke. However, anticoagulation can increase the risk of bleeding and hemorrhagic stroke. Up to 7% of strokes in low and middle-income countries may be caused by underlying RHD.[50]

Bacterial endocarditis

Bacterial endocarditis is an infection on the valves of the heart. People with valves that are already scarred or damaged by RHD are more likely to develop bacterial endocarditis than people with undamaged valves. Worldwide, approximately 60% of people with bacterial endocarditis have underlying RHD.[50] People with bacterial endocarditis have fevers and the heart may be unable to pump blood effectively. It can be difficult to diagnose bacterial endocarditis and, even when it can be diagnosed, antibiotic treatment may be ineffective. Minimising the risk of bacterial endocarditis is an important part of managing RHD. The bacteria that cause bacterial endocarditis tend to come from the mouth, so good dental hygiene is an important way to minimise risk.

Atrial fibrillation

Atrial fibrillation (AF) is an abnormal heart rhythm and a complication particularly associated with mitral stenosis. People with RHD are at risk of AF because of the structural heart changes caused by RHD. AF tends to make heart failure worse, increasing shortness of breath and may causes palpitations. AF also significantly increases the risk of stroke. In endemic settings RHD is a major cause of AF.[51]

Maternal morbidity and mortality

Women with RHD are at risk of significant illness or death during pregnancy and delivery. The changes of pregnancy (increase blood volume, increased risk of blood clots, increased blood pressure and heart rate) make the heart work harder. Hearts that have been damaged by RHD may not be able to adjust to these changes and women with RHD may develop heart failure during pregnancy. The symptoms of heart failure may be confused with symptoms of late pregnancy and go untreated, increasing the risk cardiovascular collapse and death. Women who have received heart valve surgery and metal heart valves are at risk of serious bleeding from anticoagulation medication. These medications can also affect the developing baby.

BASELINE COMPONENTS

Successful RHD control programmes are comprehensive and necessarily encompass more than the delivery of clinical care. Control programmes must interact with communities, health workers and the wider health system to facilitate prevention and treatment of RF and RHD. These partnerships need to be maintained over many years before the burden of disease is significantly reduced at a population level. Long term collaborations can support health system strengthening.

The first section of TIPs addresses elements which may be overlooked amidst provision of direct clinical services; including governance, fundraising and collection of baseline epidemiologic data. Wherever possible, baseline components should be considered (but not necessarily completed) before beginning an RHD control programme. Careful attention to baseline components can simplify the administration, sustainability and monitoring of RHD control programmes over time. Systems issues remain important throughout the duration of the programme and should be reviewed, revised and strengthened as progress towards RHD control continues.

One of the most important roles of new and renewed RHD control programmes is to gather epidemiologic data to produce a burden of disease estimate. Understanding the burden of disease makes it possible to assess the importance of RF/RHD in your setting, focus interventions in the areas of greatest need and facilitate monitoring of programme impacts over time. The information is invaluable to decision makers in government, funding agencies and communities.

Programmes are best able to respond to the burden of RF and RHD when supported by good governance, sustained funding, human resources and a structure for evaluation. It may also be necessary to work with other parts of the health system – laboratories, training providers, primary care structures, disease notification agencies – to delivery disease altering interventions. This integrative approach ensures that RHD control is part of the routine system of healthcare delivery and can be sustained over time.

The post infectious nature of RHD creates a unique opportunity for disease control to encompass a broad range of sectors and services. Interventions span from primary care to open heart surgery, from communicable to non-communicable disease and between paediatric and adult populations. RHD exemplifies the ideal integrated, diagonal, healthcare delivery in low resources settings. Well designed and delivered programmes can become beacons of best practice for other disease communities.

In most countries the health system is roughly arranged from primary care (small, local, general) to higher levels of care (larger, specialised, centralised). However, the names, capacities and structure of these levels varies between countries. Differences in terminology make it difficult to describe the levels of the health system in a way that is meaningful to all the users of this handbook. A summary of different terms appears in Table 1. You should define and adapt your own local terms when developing local guidelines and protocols for referral between health services.

The Needs Assessment Tool (NAT) provides a template for health system assessment in your setting which can be helpful for developing a baseline description of your area.

Table 1: General levels of healthcare in low resource settings and their relevance to RF/RHD

	PRIMARY	SECONDARY	TERTIARY	QUATERNARY
TYPES OF CLINICS	Health clinics Health centres Family doctors	Local hospitals	Referral hospitals National hospitals	Specialised national or regional units Visiting services *May include international services
TYPES OF STAFF	Community healthcare workers Nurses	Nurses Doctors	Nurses Doctors Specialist doctors	Nurses Doctors Sub-specialists doctors
TYPES OF SERVICES	Medications and injections	Diagnostic facilities Admission for management of symptoms	Diagnostic tests Advanced medical cared	Advanced diagnostic services Surgical services
RELEVANCE TO RHD	Diagnosis and management of Strep A infections Identify possible RF and refer for diagnosis/ management Support access to contraception for women with RHD	Confirm diagnosis Manage symptoms Advise on follow up	Assess suitability for cardiac surgery interventions	Provide or coordinate clinical care

1. BURDEN OF DISEASE DATA

THINGS TO CONSIDER

- Who does your RHD control programme provide care for?
- How can you count and describe that population?
- What sources of burden of disease data are available?
- Can these sources be combined to provide a realistic burden of disease estimate?
- Are there vulnerable groups within your population who may have higher prevalence of RHD?

"One key challenge in addressing RHD lies in the lack of reliable data capturing the true burden of disease – i.e. precise geographical, age and gender distribution and health systems measurements such as where and when care is being sought, by whom and at what cost. Health authorities in many countries rely on regional estimates of the burden of RHD given the absence of national disease registries and underreporting or misdiagnosing of acute and chronic cases of RHD."

Dr Agnes Binagwaho, Minister of Health, Rwanda, 2013[52]

Burden of disease background

'Burden of disease' is a broad term generally used to mean the number of people living with RF/RHD or dying from the disease. Burden of disease data is important for advocacy, planning and delivery of successful disease control programmes. Data is important to:

- Decide whether RF/RHD is a public health priority.
- Provide a baseline to identify targets and monitor the impact of any intervention.
- Motivate governments and funding organisations to engage with your programme.
- Understand how clinical tests, tools and guidelines will perform in your setting.

In most cases the best way to start to determine the burden of disease is with a systematic review of what is already known about Strep A throat infection, RF and RHD in your country. A systematic review can include informal 'grey' literature and will identify any existing studies. This provides a firm foundation for further research or data collection. A systematic review protocol is published in the Needs Assessment Tool to help structure your search. Access to article databases Pubmed and Embase is required to use the protocol. If you do not have access to peer reviewed journal databases, in some circumstances, RHD Action may be able to provide literature review support. Please contact us at info@rhdaction.org if required.

In some places, a systematic review will reveal that there is no published data on Strep A, RF or RHD in your setting. In this case it may be necessary to collect information to better understand the burden of disease and engage others. Published literature from nearby countries may prove informative in some cases. Finally, the Institute for Health Metrics and Evaluation provides modelled estimates of RHD prevalence and mortality for most countries including in those countries lacking primary data. While these figures are no substitute for collecting primary data, they can be a useful starting point for discussions and advocacy.

The importance of denominators

Knowing how many people your programme delivers care for is important when interpreting the burden of disease. For example, your RHD control programme may be focused on:

- A specific geographic area.
- A specific sub-population.
- A specific age group.
- A combination of the above.

The total number of people in the catchment area of your programme is the denominator, allowing you to calculate the prevalence of RHD. Understanding the total number of people you provide care for is also important for monitoring trends over time. If your population changes – through growth, immigration or rezoning – it may mask changes in the burden of RF and RHD. Denominator data may come from a census or estimates from non-government organisations (NGOs). Identifying and documenting your denominator should occur before burden of disease calculations begin.[53]

BOX 1:
Burden of disease terms

When comparing incidence (or prevalence) between two groups it is important to consider the relative size of the groups – the denominator.

PREVALENCE

The number of cases of a disease in a population at a point in time. The prevalence of RHD is usually expressed as xx/100,000 or x/1000 at a point in time. RF is a relatively short illness (usually a matter of weeks) so measures of 'prevalence' are generally not helpful.

INCIDENCE

The number of new cases of a disease in a population over a period of time. The incidence of RF is usually expressed as the number of RF cases per year per population. RHD usually begins with a long latent period so it is difficult, and often not practically helpful, to estimate the incidence of RHD.

Sources of burden of disease data

Multiple sources of information can provide an indication about the burden of RF and RHD. These sources may need to be combined to provide a 'best guess' estimate of the burden of disease in your setting. The estimate can be refined over time as more information becomes available.

In most places, reviewing clinical records provides a useful foundation for understanding burden of disease. The best quality data is collected prospectively, allowing for focused collection of information about primary care presentations for sore throat, RF, RHD, complications of RHD and pregnancy outcomes. A template for this kind of Clinical Record Review is provided in the Needs Assessment Tool and offers a rigorous approach to understanding RHD burden in a defined geographic location. This approach usually requires some commitment of financial and human resources and ethics approval for research.

Smaller, retrospective, data collection may be possible before undertaking comprehensive NAT assessments. Other indirect sources of RHD disease burden may also be identifiable. These alternative sources are outlined in the following sections.

1. Clinical record review

Hospital or health records

Many hospitals record the admission or discharge diagnosis of inpatients. Reviewing these records can provide a guide to the number of cases of RF and RHD in a community. Hospital data tends to underestimate disease prevalence because only people who present to health services, and are admitted, will be recorded – potentially missing people who are unable to access healthcare, or who have symptoms that are too mild to seek medical aid. Alternatively, tertiary or specialist hospitals often accept patients from a larger geographic area than local health services. It is important to know what health facilities serve your target population and whether their catchment area(s) include populations outside your programme.

In some places injections delivered by a health centre or hospital are recorded in an 'injection book' or a 'BPG Register'.[54] By using these books or other records of care delivery, it may be possible to identify people with RHD who are already receiving secondary prophylaxis antibiotic injections. This can provide a clue to the baseline level of disease but will significantly underestimate prevalence because only people who have been diagnosed and are receiving treatment will be included.

In places with echocardiography services it may be possible to audit the results of echocardiograms over a period of time to get an indication of health service use for RHD. For example, in Zimbabwe a retrospective review of 308 echocardiograms in 2012 revealed that 16% of people in the case series had RHD.[55] Similarly, an audit of the results from 1130 first paediatric echocardiograms in Cameroon revealed 5.8% had definite RHD.[56] In Uganda evaluation of 500 consecutive scans revealed 11% of people requiring echocardiogram had RHD.[57] Although this 'echo audit' methodology can't be used to calculate population prevalence of disease it can provide an indication of the proportion of health service utilisation that is associated with RHD.

Death records

Details about the number and causes of death – sometimes called vital statistics or mortality records – are collected in some countries. These records can provide valuable information about the burden of RHD. However, vital statistics have incomplete coverage of populations in low- and middle-income countries. People from remote or Indigenous populations, where the risk of RHD is the greatest, may also be under-represented. Even when mortality data is collected, RHD may not recorded as the underlying cause of death. For example, an audit of mortality data in Western Australia showed that one third of deaths from RHD had been attributed to other causes of death.[58] A fundamental challenge is that the accepted international system of death certification only allows for one underlying cause of death. For instance, a person with mitral stenosis who develops atrial fibrillation, then has a fatal ischaemic stroke, may be coded as dying from RHD, atrial fibrillation, or stroke. When analysing death records, it may be helpful to solicit information on local physician death certification practices in order to put these data in context. In some places, autopsy data may provide information about the burden of RHD, though autopsies are not usually performed on representative samples of the population.[59,60]

Disease notification data

In some places RF is a notifiable condition, providing valuable information about the rate of disease over a period of time (see Box 1). However, under-reporting is common, particularly when systems to report cases are weak. As RF and RHD are largely a clinical diagnosis, notifications are susceptible to change following education or outreach activities. For example, increased awareness of diagnostic criteria and case detection may lead to an increase in RF notifications and the overall incidence in RF. See Chapter 7 for details on the role of RF notifications.

2. Focused data collection

Clinical record audits

Detailed clinical audits can be performed to provide more information about the local burden of RHD and progression over time. The usefulness of these audits depends on the original data collected – for example, how well the diagnosis of RHD was recorded and whether it was confirmed with echocardiography. In some settings it may be possible to link audit data with outcome measures, including mortality. For example, in the small Pacific Island of Wallis a single electronic computer system made it possible to extract all people with a recorded diagnosis of RF and RHD over an 8 year period. These records were linked to echocardiographic reports and surgical information to provide a comprehensive overview of RHD in Wallis.[61]

3. Modelled estimates of the burden of disease

Global burden of disease data

The Global Burden of Disease Study (GBD) is a worldwide consortium working to document causes of premature death and disability of over 300 diseases in 195 countries.[62] Using a standardised approach, GBD makes it possible to track changes in disease burden over time. Summaries of GBD data are published regularly to provide updates on these changes.[63] Disease specific publications are also released periodically. In 2017 the GBD RHD Working Group produced the most comprehensive update on the global burden of RHD.[2] This paper estimates the prevalence of RHD, deaths from RHD, and summary measures of health like disability adjusted life years (DALYs) from RHD for 195 countries and territories. It does not include data on Strep A throat infections or RF.

It may be possible to view details about the modelled estimate of RHD in your country using data visualisation tools from the GBD study. These are available online at https://vizhub.healthdata.org/gbd-compare/ and further technical support is available by contacting info@rhdaction.org if required.

Extrapolating from similar countries

Even without local data it may be possible to estimate the burden of disease from similar areas or countries. This is the approach of the Global Burden of Disease project using formal statistical models. Settings with similar economic development are likely to provide the best estimate.

Historic estimates

Before the widespread use of echocardiography, cardiac auscultation was commonly used to screen schoolchildren for heart murmurs and RHD.[64] Although auscultation is now known to underestimate the true burden of RHD, historical studies provide some information about clinically significant disease prevalence.[65]

Auscultation without echocardiographic confirmation is no longer considered an appropriate approach to screening for RHD and new projects of this kind should not be initiated. See Box 24, Chapter 20, Echocardiographic screening.

4. Echocardiographic screening

Population echocardiographic screening is the current gold standard for estimating the prevalence of RHD. Rigorously conducted echocardiographic screening can provide an important burden of disease baseline for new control programmes. However, the role of echocardiography in clinical management of disease is still under investigation and not currently recommended.[66] The risks and benefits of echocardiographic screening are outlined in Chapter 20.

Opportunities for integration

Vital statistics registers are essential elements of a health system. Improving mortality data allows for improved services for a wide range of conditions. In countries with weak mortality reporting infrastructure, RHD control programmes may provide an opportunity for improving data collection. Interoperability between systems is an important consideration to ensure communication between multiple data sources. Resources and information about strengthening vital statistic registers are available from WHO.[67]

Burden of disease in specific populations

The burden of RF and RHD varies between, and within, populations and across time. This means that subgroups within your community may have a higher prevalence of RHD than others. RHD is most common in vulnerable groups including Indigenous communities and socially and economically disadvantaged people.

Burden of disease varies by age

RF and RHD have a relatively predictable age distribution worldwide, illustrated in Figure 11. RF typically occurs in people aged between 5 and 20 years, with a peak incidence of first episode of RF at 11–12 years. Symptomatic RHD can begin in childhood and prevalence increases with age.[50]

The age distribution of RF and RHD is important for estimating your local burden of disease. Cases of RF and RHD in school children may be more likely to be diagnosed (through screening or school health programmes) but they represent only 15–20% of total cases.[68] The all-age prevalence of RHD is expected to be 5–7 times higher than the prevalence in 5–14 year olds.[50] Developing countries with a high burden of RF also have very young populations which should be taken into account when reporting on the disease burden, especially if trying to compare with other countries. There are statistical techniques to do this (e.g. age standardisation), but the simplest way is to present a breakdown of RF incidence or RHD prevalence in age stratified blocks (ideally, in 5-year increments, e.g. 5–9, 10–14, 15–19 year olds, etc.) to reflect the variation in risk with age.[53]

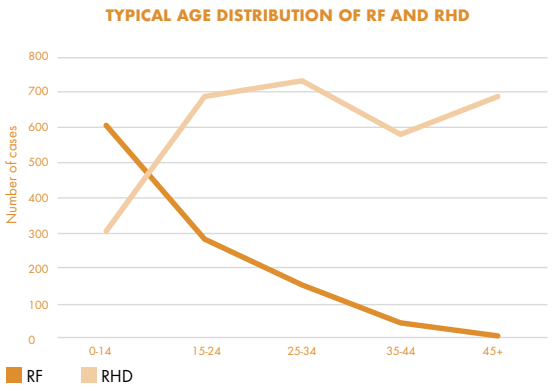


Figure 11: Typical age distribution of RF and RHD. Australian Institute of Health and Welfare.[69]

Indigenous populations

Globally, RF and RHD are more common in Indigenous communities than non-Indigenous communities.[70,71] This association has been demonstrated worldwide:

- In Canada, the population incidence of RF is 2.9 cases per million.[72] Yet in First Nations communities the incidence of RF is 75 times higher at 21.3 cases per 100,000 people.[73,74]
- In the United States, episodes of RF in the 1990s predominantly affected low income African American inner-city children.[75] Similarly, a six year retrospective study from Chicago demonstrated that 33% of people presenting for care for RHD had immigrated from outside the United States. Ninety-eight percent of the remaining people with RHD were African American.[75,76] In Hawai'i the disease is far more common in Polynesian children.[77,78]
- In New Caledonia, an echocardiography screening study showed that children of Melanesian descent had a far higher prevalence of RHD (13.5/1000) than children of European descent (1.8/1000).[79]
- In Australia, RF/RHD affect Indigenous communities almost exclusively: 97.6% of first episode RF between 1997 and 2010 occurred in Aboriginal and Torres Strait Islander Australians.[80] Indigenous Australians are 122 times more likely to live with RHD than non-Indigenous Australians.[43]
- In New Zealand, the vast majority of people diagnosed with RF are Māori or Pacific peoples.[81,82]

The high burden of disease in Indigenous populations probably stems from socioeconomic inequality. Although there are some genetic associations with RF and RHD, genetic predisposition is unlikely to explain the consistent association between economic vulnerability and burden of disease.[83]

Variability in distribution of disease can complicate burden of disease estimates. Relying on data from only one location in a country may give a misleading picture of prevalence or incidence. You may also need to consider the following points to address the needs of vulnerable populations:

- Your programme should attempt to collect sufficient and appropriate demographic detail to identify groups experiencing a greater burden of RF and RHD.
- Programme planning and activities should reflect the needs of vulnerable populations with a high burden of RF and RHD. Identifying these communities, their representatives and distribution should be addressed during the collection of burden of disease data.

Refugees and migrants

The global refugee crisis is contributing to the burden of RHD and changes in the distribution of disease. As of 2015, 65.3 million people around the world have been forced from their homes.[84] Of these are 21.3 million refugees, over half of whom are under the age of 18 years, when the risk of RF is greatest.

More than half of all refugees come from the Syrian Arab Republic, Afghanistan and Somalia.[84] Poverty and inadequate and disrupted primary healthcare systems are major contributors to the persistence and resurgence of RF/RHD in source countries. Prior to the conflict in Syria, RHD was the leading indication for valvular heart surgery.[85] Displaced Syrian children continue to present for RHD surgery in neighbouring Jordan.[86] In Afghanistan, there are case reports of high burden and unmet need for humanitarian evacuation for cardiac surgery.[87] Médecins Sans Frontières has been using echocardiography to screen refugees aged 10–25 years for RHD in Rome, Italy, and have identified a high burden of disease.[88]

Recently arrived migrants, refugees and displaced persons from settings affected by crisis often have complex health needs. Many migrants experience difficulties in continuity of healthcare and record keeping. Some refugees lack access to any health records, continuity of service or provision for chronic disease. This makes identifying and managing RHD particularly difficult. For example, following the breakdown of the Union of Soviet Socialist Republics (USSR) RHD prevalence increased significantly in association with social and economic disruption.[89] Similarly, in Nigeria and Côte d'Ivoire political instability has disrupted access to echocardiographic diagnosis of RHD.[6]

The Strep A infections which cause RF and subsequent RHD spread most easily in overcrowded communities. Worldwide, refugee settlements are grossly overcrowded fostering conditions for the spread of RHD.

RHD is rare in developed countries, outside some vulnerable Indigenous populations. This means that doctors and healthcare workers in resource rich settings may not be familiar with RHD, leading to late diagnosis and poor care. The infrastructure for delivering register-based secondary prophylaxis is often not in place in developed countries. In the United Kingdom, maternal deaths in recent immigrants with RHD highlights the challenges of providing comprehensive care in settings where the disease is now rare.[90] There have been strong calls for receiving countries to strengthen care capacity of refugee children with RHD.[91]

2. GOVERNANCE

THINGS TO CONSIDER

- How will decisions be made about your RHD control programme?
- Does your RF/RHD Advisory Committee include representation from all key stakeholders?
- Are people living with RHD represented on your committee?
- Does the committee have a clear role, timeframe and terms of reference?
- What are the primary goals for the programme?

Designing, developing and sustaining an RHD control programme requires input from many different stakeholders. It can be helpful for the goals, strategy and planning of the programme to be overseen by a committee to represent these different group. Having an established governance mechanism may also be useful when decisions need to be made throughout the programme to inform evaluation and scale up.

A group of people with a mandate to focus on RHD can also advocate for prioritising the disease – even when comprehensive programmes are not yet established. For example:

- In India a 'National Rheumatic Heart Consortium' was established in 2011 to formulate national guidelines and advocate to the government on issues related to RHD.[92]
- In Canada an 'ad hoc Acute Rheumatic Heart Disease Working Group' was formed in 2015 to respond to an increase in RF cases, focusing on provision of treatment and investigation of the increased RF burden. Membership of the Canadian working group including Government representatives, the First Nations Health Authority, local health clinic and local research organisations.[93]
- In Australia an END RHD Coalition has been formed to support research efforts towards an endgame for RHD in the country.[94]
- In Namibia a 'National Advisory Committee on Rheumatic Fever and Rheumatic Heart Disease' has been established to develop an RHD control programme in conjunction with the Ministry of Health.[95]

Committee membership

A diverse advisory committee offers the best opportunity to address the primary needs of each stakeholder. Potential membership is outlined in Table 2.

"The programme advisory committee consisted of a part-time provincial cardiologist, supported by representatives of related departments, such as paediatrics, cardiology, primary healthcare, hospital care and epidemiology, as well as microbiology laboratories, nurses and a representative of the Ministry of Education. There was one collaborating physician as a local representative in each hospital."

Nordet et al, Prevention and control of rheumatic fever and rheumatic heat disease: the Cuban experience, 2008[33]

Table 2: Potential representatives of the RF/RHD Advisory committee

GOVERNMENT*	HEALTH SECTOR	COMMUNITY	NON-GOVERNMENT
Ministry of Health Ministry of Education Ministry of Housing *Ministry of Indigenous Affairs where relevant	Primary care/general practice/family medicine Community health workers Nurses Midwives Traditional healers Public health physicians Cardiologists Paediatricians Internal medicine physicians Cardiac surgeons or representatives of surgical programmes Dentists Epidemiologists Microbiologists Laboratory services Echocardiographers	People living with RHD Community leaders Faith-based groups Family groups Schools and teachers	WHO Donors and funders National/international Heart networks and societies Private sector partners International technical advisers Academic or research institutions

Roles and activities of the Advisory Committee

Function of the RHD Advisory Committee

The goals of the committee should be clearly articulated in the Terms of Reference (ToR). The ToR may need to change over time as new priorities and challenges emerge. The ToR needs to be clear about the purpose and role of the committee, the extent of its decision-making abilities and expectations for meetings. It may also specify how long the committee will function before evaluating its effectiveness. Planning for programme evaluation is discussed further in Chapter 10. Many disease control programmes have an advisory committee and it may be possible to identify similar committees in your area with a ToR suitable to adapt to your programme needs.

For example, the goals and function of the advisory committee in Western Australia (WA) were described in evaluation as: 'The WA Rheumatic Heart Disease Clinical Advisory Group provides advice and guidance supporting clinical functions and interactions of the WA RHD Programme. This group helps draft WA action plans, monitoring their implementation by the WA control programme and evaluating impact. Meeting quarterly, membership includes paediatric and adult cardiologists, primary health care providers, Aboriginal Health Council of Western Australia (AHCWA), Aboriginal Medical Services, ARF/RHD Experts, WA Communicable Diseases Control Directorate and West Australian Country Health Services (WACHS).'[96]

Subcommittees may need to be formed to focus on specific pieces of work, particularly adapting clinical guidelines to local settings. Administrative support to arrange meetings, book venues, record minutes and follow up on action items is also very helpful. You may be able to include this kind of administrative support in your budget and fundraising proposals.

Potential roles of the RHD Advisory Committee

The 'Policy and clinical context' modules of the RHD Action Needs Assessment Tool provides a good starting point to get an overview of current RHD activities, stakeholders and priorities. An interview guide is downloadable online and may inform your planning activities. An overview of the potential roles of the RF/RHD Advisory Committee can be found in Table 3.

Table 3: Potential roles for the RF/RHD Advisory Committee

CLINICAL LEADERSHIP	Develop evidence-based, locally adapted, clinical management guidelines. Develop consensus about management plans to standardise patient care, and provide clarity for clinicians at each level of the health system. Support excellence in care delivery.
SETTING STRATEGY AND GOALS	Identify gaps in local data, gather or support research to fill them. Identify goals and targets for the programme. Tracking activities and data to guide the priorities of the control programme staff.
REPRESENTING THE PROGRAMME AND BUILDING RELATIONSHIPS	Present the work of the programme at meetings, in the media and to the community.
ENGAGING THE COMMUNITY AND OTHER STAKEHOLDERS	Represent the programme to people living with RHD, including RHD clubs and advocacy groups. This representation makes it possible to tailor programme activities to best meet the needs of individuals, families and communities. Foster relationships with other professional organisations in the country.
RESOURCE MOBILISATION	Fundraise, mobilise resources or advocate for financial support of the programme.
ADVICE AND MENTORSHIP	Support individuals and programmes to expand RHD control activities in other locations.

The critical role of people living with RHD

There is increasing global recognition that people living with disease should be included in priority setting and decision making about how that disease is addressed. This is true for individual clinical decisions and equally true for larger policy choices and research projects. Full participation in decision making is often summarised as 'nothing about us without us'.[97] This participatory approach is empowering, respectful and ensures that choices best reflect the needs of ultimate end users.[98]

"It's about your experience, you've had the operation, you've suffered with the condition, you've had the treatment...

Sometimes the doctors are working by the book and what they see (in the exam room) but they don't really know what the patient is experiencing. It could be anxiety because you don't understand, you think you're going to die or worried your quality of life might be affected...

In lots of cases, the doctors talk over your head – they're conferring with one another. You're sitting in that room but you're not in that conversation. When I'm in the clinic, I'm not part of the conversation, the doctor doesn't hear me (as a whole person); but when I'm in the advisory group my conversation is heard and it is important."

RHD Patient Community Advisory Group Member

Local leadership in control programmes

New RHD control programmes are sometimes supported by international partners. These may include international governments, academic institutions, hospitals or funding agencies. In these cases, a strong advisory committee is a vital mechanism for ensuring the programme has local governance. For example, the 2016 All-Africa Workshop on RF and RHD advocated for the voice of the 'front-line African health leader' to be integral in priority setting for this disease.[99] International support is generally for a fixed period of time. It is essential that programmes develop a sustainability plan to continue work beyond the end of international support.

"From the very outset, it was understood that the success of local programmes would ultimately depend on the extent to which countries themselves are able to maintain the program activities as part of their national health services."

WHO Global Programme for the prevention of rheumatic fever/rheumatic heart disease in sixteen developing countries. Meeting of national programme managers, Geneva, 4–6 November 1986.[100]

3. FUNDING

THINGS TO CONSIDER

- Do you have sufficient funding for your programme or activity?
- Do you have a fundraising strategy for your programme or activity?
- Do you have evidence that supports the need for funding?
- How will you recognise or acknowledge charitable donations to your programme?
- Will you provide fact sheets or resources to individuals, families or communities who wish to undertake their own fundraising?
- Do you have a strategy for dealing with any conflict of interest?

The WHF Roadmap for RHD Control highlights that programmes require sustained long-term funding to realise population-level impact, and therefore rely on strong advocacy and a good understanding of programme costs.[101] The need for ongoing funding reflects the protracted causal pathway from Strep A infection to RF and RHD, spanning decades. The brief case studies at the start of this resource demonstrate that comprehensive control programmes generally take a decade before they are able to show evidence of a reduced disease burden. Interim measures of programmatic success are discussed in Chapter 10.

Ideally, sustained, predictable funding for RHD control activities will be provided by governments. This reflects the evidence that RHD control is cost effective and good value for money.[102,103] However, securing substantial funding for initiating new programmes is relatively rare. More commonly, RHD programmes begin with a patchwork of smaller funding sources – for example for one-off activities such as nurse training or development of clinical guidelines – which can grow and coalesce into a unified programme as relationships develop and it is possible to demonstrate success. This 'patchwork' approach has underpinned some large RHD initiatives worldwide, including the programme in Fiji outlined in Box 2.

BOX 2

CASE STUDY MIXED FUNDING FOR RHD CONTROL IN FIJI

The RHD Programme in Fiji has benefitted from a variety of innovative approaches and partnerships. Since 2009, Cure Kids New Zealand has supported RHD activities in Fiji through ongoing funding from foundation key partner Accor Hotels and support from the Fiji Water Foundation. Between 2014 and 2019 the New Zealand Partnerships for International Development Fund, together with Cure Kids New Zealand and technical partners, was a major contributor toward the rapid expansion of the Fiji RHD Control and Prevention programme.[104] This included a formal partnership with the Fiji Ministry of Health and Medical Services, specific funding for recruitment of an eight member team, development of a web-enabled rheumatic fever information system. With more than 10 years of research and foundational work, the national technical advisory committee and the Ministry were well positioned to attract large scale funding for this scale up phase.

Vicki Lee (former CEO Cure Kids NZ), Mr Jone Usmate (former Minister for Health, Republic of Fiji) and Helen Leslie (Ministry of Foreign Affairs and Trade, New Zealand Government representative) celebrating the launch of the next phase in the RHD prevention and control programme in Fiji.

Preparing for finding finance

In order to initiate or expand RHD activities most control programmes seek funding from a variety of sources. Whether a single or multi donor approach it is important to clearly describe the activity, timing and duration, and what resources, in kind or otherwise, are needed. This will need to be itemised for the donor. Where there are other contributors, such as in-kind contributions from the Ministry of Health or volunteer time it will be important to highlight this in the proposal so that the potential donor can have as full a picture of the activity as possible. Considering these issues in advance will make it easier to apply for additional funding as opportunities arise.

Develop a programme or activity plan

A programme plan with an accompanying budget demonstrates a readiness for action and may increase the likelihood of obtaining external funding or budget allocation. Forming an advisory committee, as outlined in Chapter 2, demonstrates the engagement of key stakeholders and can provide valuable planning input. You should include estimated burden of disease data, rationale for the planned activity, and activities you propose to conduct along with associated targets (for example, indicative number of trainings and number of beneficiaries or participants). Where possible, ensure that the plan and outcomes are aligned with local integrated health plans, annual corporate plans or a national non-communicable disease (NCD) action plan.

Develop a budget

A carefully developed budget may help build confidence and trust between the potential funders and programme implementers. It will also help ensure that the amount of funds being requested reflect what is needed to conduct the activity. Burden of disease data, for example, may be helpful for estimating the number of people living with RF and RHD, which may also help you estimate staffing requirements, medication, transport and other costs. Many government and non-government funding agencies and donors have specific requirements for budget preparation, so the budget may need to be revised for each specific application. It can be helpful to have an independent peer or colleague review your funding application. You may consider approaching colleagues running similar control programmes, Ministry of Health officials or other international organisations who offer specialised technical support to help review your budget.

Explaining why funding is needed

Document the existing costs of RHD

It is common for countries to use considerable resources managing advanced RHD without necessarily appreciating the costs of disease. In settings with a high burden of RHD many people present with advanced heart disease and require specialist care. Often this cost is paid by the Government or the health services. Health service spending on admissions and heart surgeries often means that very little money is being spent on prevention. It can be helpful to collect data about the cost of RHD to demonstrate the scale of the problem and to motivate action. The costs associated with RHD led to primary prevention of RF and secondary prophylaxis to both be identified as 'best buys' for tackling heart disease, diabetes and stroke in the African region.[102]

Analyse the cost and benefit of potential interventions

Choosing the right mix of primary, prevention and tertiary interventions to care for people currently living with RHD – and to minimise the development of new cases – is challenging. A decision-making tool has been developed to help policymakers address this question.[103] The tool uses an economic model to estimate the impact of scaling up primary prevention, secondary prevention or heart valve surgery. By inputting local data it is possible to estimate the incremental cost effectiveness ratio (ICER) of each option. The tool has been developed to support countries in Africa in particular – but its principles are broadly globally applicable. A downloadable excel file to use the tool is available online. In some cases, technical support to use the tool may be available from RHD Action – you are welcome to contact us to discuss cost effectiveness modelling projects at info@rhdaction.org

Estimating the economic impact of a control programme is sometimes called a 'business case'. Cost effectiveness analysis is a more formal approach for analysing costs and benefits of interventions. A cost effectiveness analysis can help decision makers know how to allocate limited resources. There have been a number of projects to explore the cost effectiveness of RHD control in different settings.[16,105,106]

RF and RHD are chronic conditions accruing considerable personal and social cost over many years. Some of these costs are direct and tangible; others are indirect or opportunity costs as outlined in Table 4.[107] Minimising the financial burden on individuals and identifying cost effective disease control strategies for populations is an important global goal.

Table 4: Costs of RHD

DIRECT COSTS	INDIRECT COSTS
INDIVIDUALS, FAMILIES AND COMMUNITIES	
People and families living with RHD have to spend money to manage symptomatic disease. Outpatient costs include medication, transport to appointments, dental care and blood tests. Inpatient costs may include payment for admission, laboratory tests, surgery, food and accommodation. In some countries people are responsible for almost all of their own healthcare expenses; these 'out of pocket' costs drive medical poverty and personal bankruptcy.[108]	RF and RHD are also costly through reduced social and economic participation.[107] Young people with RF or RHD may be unable to complete or excel in schooling. In West Africa children with severe RHD have poor educational outcomes.[6] Additionally, parents may need to stay home from work to care for unwell children. In Brazil, nearly one quarter of parents took time off work to attend children with RHD and nearly 5% lost their job as a result.[109] In West and Central Africa many people with RHD require long periods away from work or face termination of employment.[6] RHD also causes a 'cost' to quality of life as people with the disease worry about their future and experience symptoms. In Brazil, quality of life impairment from RHD was similar to the effect of living with other chronic conditions such as asthma and epilepsy.[110] In Egypt, 98% of parents of children living with RHD are concerned about the family and financial impacts of the disease.[111]
GOVERNMENTS AND THE HEALTH SYSTEM	
The health system cost of RF and RHD can be enormous, particularly in countries where governments subsidise or pay for healthcare. Most of the costs are incurred in tertiary treatment for severe disease, including hospitalisation and surgery.	Indirect costs occur through reduced participation of people living with RHD and their families. People who are too sick to work or who have died earlier than otherwise are not able to contribute to economic growth. Indirect social costs are often not included in routine cost effectiveness analysis – but they can be significant. In South Korea 39% of the total cost of RHD was found to be indirect, through reduced productivity and premature death.[112]

Sharing the lived experience of RHD

There are many practical, economic and ethical arguments for RHD control. These should be outlined for your setting based on locally available data and evidence. However, political and funding decisions are often influenced by personal connections, stories and experiences. Sharing the stories and experiences of people living with RHD can make the issue relatable for policy makers, engage their interest and drive motivation for action. In some settings you may find that health leaders, for example, Ministers or permanent secretaries may not come to their role with a medical or public health background. Engaging people with a personal experience, to share their story, can sometimes to help convey an advocacy message more effectively.

RHD Action has a number of 'People living with RHD profiles' and video resources which can help share some of the experiences of people living with RHD – these can be accessed online. Customised resources for specific settings are particularly impactful. Identifying people living with, or impacted by, RHD in your setting means local stories can be shared with local decision makers – either by attending meetings, developing patient profiles, media interest or video footage. Ideal RHD advocates are confident public speakers with a good understanding of the disease.

Sources of funding

Globally, many different kinds of organisations, private and not for profit, have funded RHD control programme activities, equipment, events and advocacy. An overview of potential funding groups is presented below. This is not an exhaustive list and novel opportunities for funding should be explored wherever possible.

Development assistance: Some governments have funded international control programmes and humanitarian surgical missions. Often these funds are arranged at a government level and require diplomatic collaboration. Some funding may also be available through WHO Country Offices or grant programmes.

Professional organisations: Professional groups, including medical associations or professional colleges, can be instrumental in generating or helping to secure funds for supporting project activities.

Charitable donors: A variety of charities have donated funds or resources to RHD control programmes. These valuable contributions often support small local initiatives. For example, in Australia charitable groups supported development of an RHD video which was widely used throughout the Northern Territory.[113] Community organisations, faith-based groups, business foundations and NGOs may prove to be valuable partners for your programme.

Businesses: Local (or international) businesses can sometimes be encouraged to donate funds, often for specific pieces of equipment. Businesses may also be willing to contribute their own products – perhaps including meals for people attending RHD clinics or meetings, pens or paper to clinics, paint for education campaigns or other services. See Box 3 for additional information on donations of medical equipment.

Community fundraising: In many countries, families or individuals seek private support for cardiac surgical interventions. Fundraising generally happens with events, through media appeals or via social media. You should consider how your programme will respond to these efforts. Personal appeals may be a good opportunity to raise awareness about RF and RHD but it can be difficult to ensure that information is accurate and appropriate.

Research funding: Appropriately, many RHD research projects in low resources settings have a service delivery component. Any research conducted in conjunction with your programme should address the clinical needs of people and communities living with RHD.

"Medical research involving a disadvantaged or vulnerable population or community is only justified if the research is responsive to the health needs and priorities of this population or community and if there is a reasonable likelihood that this population or community stands to benefit from the results of the research."

Declaration of Helsinki - Ethical Principles for Research Involving Human Subjects, 2008.[114]

International research collaborations offer considerable opportunity to share knowledge, skills and experience. However, practicalities and expectations should be carefully considered and documented from the outset. Research questions must also reflect local needs – as determined by local decision makers – and be coupled with clear plans for translating research outcomes into practical benefits.

Fundraising resources

Raising funds, maintaining relationships with donors, minimising conflict of interest (see Box 4), reporting on outcomes and building financial sustainability is a specialty field in its own right. Larger programmes should consider review of fundraising resources or expert advice.[115] A large number of fundraising toolkits and resources for other diseases are also available online and may be adapted to RHD.

BOX 3:
Medical equipment donations

Donations of medical equipment are a popular form of support for health programmes in resource limited settings. Providing tangible items offers donors an opportunity to be photographed with the product and be recognised with a plaque or an unveiling event. However, donations of medical equipment can cause unexpected problems, including the cost of maintenance, trained operators, location of the donation and inequitable access to resources.

WHO has developed an excellent resource "Medical device donations: considerations for solicitation and provision" outlining many of these considerations in detail.[116]

It can be helpful to be clear about what you need funding for the most (usually by developing a budget) and encouraging potential funders to give to the areas of your highest priority. This may help avoid excessive investment in equipment, over training or staffing. Having a plan for recognising donors – through events, photos, openings or public acknowledgment – may be a way to encourage funding towards intangible items, including salaries and programme costs.

BOX 4:
Conflict of interest

Conflict of interest occurs when 'an individual or organisation is involved in multiple interests, one of which could possibly corrupt the motivation for an act in the other'.[117] Conflict of interest for RHD control programmes could include relationships with donors or funding partners who have a financial interest in clinical decisions; for example, manufacturers of medical or diagnostic machines may sway a programme towards tertiary interventions, rather than primary or secondary care. You should consider how your programme can minimise these risks. More broadly, you may also consider how your programme will engage with manufacturers of health harming products, potentially including tobacco companies and companies making unhealthy food products.

4. LABORATORY SERVICES AND MEDICAL EQUIPMENT

THINGS TO CONSIDER

- Does your programme have access to a local laboratory?
- Does your programme have access to a reference laboratory?
- How do you ensure that test results are reported to the appropriate clinician?
- Do you have robust procurement systems to order and distribute laboratory resources?
- Does your laboratory have quality assurance or quality control measures in place?

While access to laboratory services is a valuable component of RHD control programmes, successful programmes have been possible with very little laboratory support. It is not essential that complex laboratory services are perfected before disease control activities begin.

The development of high quality laboratory services in developing countries tends to reflect the burden of disease in that country and, increasingly, by the global health security agenda.[118-120] A number of initiatives are underway to increase laboratory capacity in RHD-endemic settings, including the World Health Organization Regional Office for Africa (WHO-AFRO) Stepwise Laboratory (Quality) Improvement Process Towards Accreditation (SLIPTA) programme.[121] Advocacy by RHD programmes to strengthen laboratory services can occur concurrently with other interventions to address RF and RHD.

Margaret Baekimia analysing a blood sample in the Pathology laboratory at the Kilu'ufi hospital, Solomon Islands.
Credit: Rob Maccoll / DFAT

This chapter provides some background to laboratory services and outlines some of the relevant laboratory services relevant to Strep A infections, RF and RHD. Additional detailed information about laboratory procedures is available from the WHO Manual for the Diagnosis of Group A Streptococcal Infections (1996). The Manual is downloadable online. Although some parts of this resource are out of date many of the basic techniques for Strep A identification remain relevant. More contemporary manuals for Strep identification in higher resource settings have also been developed by the CDC Streptococcus reference laboratory.[122] General information on microbiology procedures in low resource settings is also available through WHO publications.[123]

A baseline assessment of laboratory services can be conducted using the RHD Action Needs Assessment Tool template. This survey can provide information about laboratory tests available and barriers to use of laboratory services.

Opportunities for integration

The principles and practices of laboratory bacteriology are not specific to Strep A. Establishing basic bacteriology facilities will greatly facilitate the clinical management of many patients with infections. In particular, basic knowledge of aseptic techniques, centrifugation, staining and microscopy alongside culture facilities to identify Strep A are valuable in a range of presentations including invasive disease.

Laboratory facilities can facilitate RF and RHD control programmes at a number of levels as outlined in Table 5.

Table 5: The role of laboratory services at different levels of RHD control

PRIMARY PREVENTION OF RF Supporting the diagnosis of primary Strep A infection for primary prevention. Discussed in further detail in Chapter 12, sore throat guidelines.	**Bacterial culture** Bacterial isolates from swabs of the throat and skin can be grown in the laboratory to detect the presence of Strep A. This generally requires inoculation of blood agar plates and incubation overnight within a fixed temperature range. Visual screening of plates enables detection of b-haemolytic colonies displaying a typical Strep A morphology. Additional testing may be used to confirm the presence of Strep A colonies such as a catalase test, Pyrrolidonyl Arylamidase (PYR) spot test, bacitracin susceptibility and a simple gram stain to observe Gram-positive cocci, arranged in chains. Commercially available Lancefield antigen grouping era can be used to differentiate ß-haemolytic streptococci.[124] Automated systems do exist but are not in widespread use in endemic settings. Persistent Strep A sensitivity to penicillin generally means than antibiotic sensitivity testing is not required prior to treatment.[124] **Rapid antigen detection tests (RADT)** RADT tests are easy to use, commercial kits that detect specific parts of the Strep A bacteria (antigens). RADT are generally used at point of care and do not require laboratory support. The specificity of rapid antigen tests is generally high however the sensitivity can vary.[124] The role of RADT in diagnosis of Strep A throat infections is discussed further in Chapter 12.
THE ROLE OF THE LABORATORY IN DIAGNOSIS OF RF Evidence of preceding Strep A infection	**Evidence of preceding Strep A infection** Strep A infection results in the production of antibodies specific to antigens expressed by Strep A. Antibody responses usually peak 3–4 weeks after infection and stay elevated for 2–3 months. The specific antibodies that are most commonly tested for are anti-streptolysin O titre (ASOT) and anti-deoxyribonuclease-B (ADB) antibodies. ASOT is a relatively low cost test which can be readily conducted in the laboratory using classic titre techniques. Newer methods are available but not in widespread use.[124] However, ASOT can be difficult to interpret clinically, particularly because the normal range changes with age and the normal range has not been defined for all populations.[125] A single elevated ASOT is often used for the diagnostic algorithm for RF. Confirmation of recent Strep A infection is more accurately obtained by sequential ASOT samples that demonstrate an increase in antibody titre. Both tests are commercially available but are often not accessible in developing countries with the highest burden of RHD. **Tests for inflammation and/or infection** Acute phase reactants include erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). There is variable elevation during the acute phase of RF with arthritis or carditis. They may, however, be normal when chorea is the only manifestation of RF. Some point-of-care CRP tests are manufactured but not yet widely available in low resource settings.[126]
THE ROLE OF THE LABORATORY IN MANAGEMENT OF RHD AND PRE-OPERATIVE EVALUATION	**Pre-operative evaluation** Laboratory tests for potential surgical candidates may include a full blood count, coagulation screen, liver function test, creatinine, glucose, urea and electrolytes.[127] Screening for infectious diseases may include tuberculosis, HIV, hepatitis B, hepatitis C and malaria. Women with RHD also require access to pregnancy tests. Further details about pre-operative evaluation appear in Chapter 23. **Anticoagulation** International Normalised Ratio (INR) is a measure of therapeutic effect from the oral vitamin K antagonist medications, including warfarin. INR facilities are essential for programmes caring for people who have received mechanical heart valve replacement and others. Details about anticoagulation monitoring appear in Chapter 22.

EPIDEMIOLOGY	Strep A are divided into different types (strains) according to different proteins on the surface of the bacteria. There are many different types of these T and M surface proteins, which are formed by differences in the emm gene of the bacteria. Strain typing can be performed at some local laboratories based on protein phenotyping. emm gene typing and clustering is used in specialist laboratories to classify different Strep A strains.[128] In endemic settings these are generally not clinically significant and are mainly used for research. Rarely, strain analysis may be a useful addition to an outbreak investigation but this should be conducted in conjunction with a reference laboratory.[129,130] Studies of Strep A strains circulating in different communities will be needed to inform progress towards Strep A vaccine development. Some strain collection registers already exist, often including samples of invasive forms of Strep A infection.[131] As progress towards a Strep A vaccine continues, RHD control programmes may have a role in collecting this data – see Chapter 15 for a discussion of RHD programme engagement in vaccine development.

Location of laboratories

Laboratory services parallel the levels of healthcare service (see Table 1) – different tests can be conducted at different locations. These can be considered in three broad categories: point of care tests, local laboratories and reference laboratories.

Table 6: Location of laboratory services relevant for different levels of RHD control

POINT OF CARE	The point of care tests most relevant to RHD are rapid antigen detection tests (RADT) for detecting Strep A infection and point of care INR testing for monitoring anticoagulation. Access to these tests in primary care is currently low in endemic settings and a wide range of systems issues impact on uptake of new technology.[132]
LOCAL LABORATORIES	Local laboratories generally provide simple diagnostic tests for routine clinical use. For example, in South Africa a standard 'Primary Care Laboratory Handbook' outlines a selection of tests which should be readily available, who can order them and how to collect basic samples.[133] These accessible tests potentially include, culture of swabs and blood tests of inflammatory markers to support the diagnosis of RF. Ideally, local laboratories are located close enough to health facilities that specimens can be transported quickly from bedside to testing facility. Delay in getting specimens to a laboratory makes it more likely that samples will degrade and results will be less accurate. Refrigeration is helpful if transport time is prolonged.
REFERENCE LABORATORIES	Reference laboratories can provide critical support to local laboratories for epidemiologic assessment or outbreak investigations.[124] Other roles may include: • Providing confirmatory testing and supply of reference strains of Strep A to ensure accurate, reliable quality controlled results. • Provide expert advice on laboratory standards and training. • Provide specialist knowledge on sample testing and result interpretation. • Perform molecular typing. • Provide some referral services and liaising with other national reference laboratories and public health bodies. For example the Centres for Disease Control and Prevention support a Streptococcus Laboratory which maintains a global database of Strep A *emm* subtypes.[134]

"Laboratory facilities are often the least accessible in places where the burdens of streptococcal disease and post infectious sequelae are highest."

McDonald et al, Recovering Streptococci from the throat in remote tropical communities, 2006[135]

Practical issues for laboratory services

Developing robust, reliable and quality assured laboratory services is a specialised technical field.[136] The particulars of establishing and maintaining a laboratory service is outside the scope of TIPs (and most RHD programmes). However, basic characteristics of quality laboratory services in low resource settings are outlined for your consideration. Accreditation systems are in place to develop quality laboratory services, including a graduated accreditation system established by the WHO-AFRO.[137]

Laboratory quality considerations in low resource settings:[138,139]

Laboratory organisation and personnel: Structure of the laboratory, leadership, management, focus on offering clinically relevant cost-effective testing and ability to identify knowledge and training gaps. Provide opportunities to educate and develop staff through bench-side supervision, collaboration and alignment with reference laboratories.

Equipment and work facilities: Improvement of laboratory infrastructure and purchasing of appropriate equipment, maintenance, servicing, calibration and documentation.

Appropriate procurement processes: Purchasing, receival, inspection, inventory, documentation and storage of resources and reagents/kits.

Laboratory Information Management Systems: Management of manual or automated patient record systems and return of accurate and reliable results to appropriate clinician in a timely fashion.

Laboratory corrective and prevention action: Identification and resolution of laboratory errors by internal audits and resolution of non-conformances.

Process improvement: Improve sample flow, traceability and turnaround time from specimen collection to availability of laboratory test result through proactive evaluation of consumers' satisfaction.

Quality assurance activities: Development of standard operating procedures (SOP's), participation in national external quality assurance (EQA) programmes, assessment of current status and working towards accreditation to international standards.

Biosafety and medical waste disposal: Safe disposal of infectious laboratory waste to minimise the potential of contamination of people or the environment.

Laboratory staff training

Formal training for laboratory staff will vary across the globe and may range from a short course to a university degree. Low and middle-income countries frequently experience shortages of experienced laboratory technicians and laboratory managers; many staff receive their training 'on the job'. This may be supplemented by a laboratory manual for Strep A to support remote training.[140] The formation of technical laboratory working groups are a great way to share ideas and build capacity in resource-constrained settings. These can come together, in a physical or virtual space (e.g. social media groups or online discussion boards) to support knowledge exchange and skill-sharing. Groups such as this provide a forum in which laboratory staff around the world can seek help, advice and information about any aspect of laboratory practice in developing locations.[141] It may be additionally useful to include laboratory staff in RHD programme education events, including workshops and conferences.

Laboratories performing microbiological testing should ensure that staff are suitably trained, and work to precise guidelines and standards to ensure that results are accurate and backed up by quality assurance programmes. Standards do not need to be complex and have been established in a wide variety of settings despite variations in resources.

Reporting of laboratory results

Samples that are being transported need to be labelled correctly and adequately so that results can be returned to the patient or ordering clinician. The return of results to clinicians and patients should occur as quickly as possible to guide clinical management. Results should be reliably documented in patient records or laboratory Information Management Systems where available. In practice, systems for recording and communicating laboratory results are often fragmented. In South Africa, half of primary care physicians indicated delayed results were a major barrier to use of laboratory services.[142] These limitations should be considered when adapting clinical guidelines for local use.

Engagement in research

Capacity building for laboratory staff can include active engagement in research projects.[143] This is exemplified by the WHO-AFRO Strep A register underway in Africa aimed towards gathering passive surveillance data about Strep A pharyngitis and active surveillance data about invasive Strep A.[131]

Medical equipment for diagnosis and management of RF and RHD

In general, medical equipment for the diagnosis and management of RF and RHD is poorly available in the settings of greatest need. In one survey, hospitals in Kenya and Uganda were asked about essential equipment and staff trained to use the devices. ECG was available in 44% of Kenyan hospitals and 28% of Ugandan hospitals. Echocardiography was available in only 28% of Kenyan hospitals and 46% of Ugandan hospitals.[144] In practice, this means that echocardiography for children in Uganda is available in only 3 private referral hospitals and a small number of private facilities.[145] This lack of diagnostic facility is common in low resource settings with a high burden of RHD.

Different medical equipment is required at each level of the health system, as outlined in Table 1. The WHO Package of Essential Noncommunicable Disease Interventions (WHO PEN) for primary care in low resource settings provides some guidance about appropriate resources for local clinics.[146] Other guidance comes from the Partners in Health Chronic Care Integration Guide for Endemic Non-Communicable Disease.[147] These resources are summarised in Table 7. Online resources are available for more specialised advice including support for procurement, stock control of consumables, sterilisation, waste disposal and maintenance of equipment.[148,149]

Table 7: Primary care equipment for RF/RHD management

RESOURCE	ROLE
STETHOSCOPE*	For auscultation of heart murmurs.
SCALES* AND TAPE MEASURE	For monitoring heart failure, nutrition and calculating body mass index (BMI).
BLOOD PRESSURE MACHINE*	Blood pressure measurements are useful for the management of a wide range of conditions and blood pressure readings are often recommended as part of in-hospital care for suspected and confirmed RF. Blood pressure measurements are also necessary for evaluating adverse drug reactions which may be associated with BPG delivery.
X-RAY	Chest x-rays can be helpful for monitoring congestive heart failure but add relatively little value than experienced clinical examination.
ECG MACHINE	An electrocardiograph (ECG or EKG) machine is needed to measure the PR interval, used in the Jones Criteria, for the diagnosis of RF. ECG is also valuable in confirming the diagnosis of arrhythmias such as atrial fibrillation.
RESUSCITATION EQUIPMENT	Including a defibrillator* and access to adrenaline for managing anaphylaxis.

*Items with an asterisk are included in the WHO PEN.[146]

Echocardiography machines

Access to echocardiography supports the clinical management of RHD.[150] One study from South Africa explored the clinical impact of introducing transthoracic echocardiography services at a district hospital.[151] The most common reason for referral to this new service was for evaluation of suspected heart valve disease. Results from echocardiography studies changed treatment or referrals plans for most patients. In general, echocardioography is valuable for:

- Investigating incidental murmurs.
- Quantifying valve disease.
- Monitoring for evidence of disease progression.
- Triaging people for operative intervention (see Chapter 23 for a discussion of pre-operative planning).
- Planning operative intervention.
- Cardiology review of valve lesion progression.
- Monitoring post-operative outcomes.

Transthoracic echocardiogram is sufficient for the majority of these tasks. Planning operative intervention may necessitate transoesophageal investigation in tertiary or specialist centres.

There is a global shortage of qualified staff to use echocardiography machines and interpret their result. Diagnostic echocardiography – as distinct from limited view screening echocardiography – requires considerable training and supervision to obtain and interpret images. Some training programmes have been developed to address this issue in low resource settings. For example, a partnership between Duke University and Moi University has amplified echocardiography capacity in Eldoret, Kenya. New echocardiography machines, capacity to store images and intensive training increased echocardiogram capacity by 149% between 2009 and 2015.[152] Some additional considerations when purchasing or accepting a donated echocardiography machine are outlined in Box 5.

Ensuring that workforce issues for echocardiography are addressed before a machine is purchased can help to avoid expensive equipment sitting idle and potentially falling into disrepair.

BOX 5:
Considerations when purchasing or accepting a donated echocardiogaphy machine

- Does it have two dimension, pulse wave and colour Doppler imaging required to apply the World Heart Federation criteria for the diagnosis of asymptomatic RHD?
- Can it store, save or transmit images?
- Is additional software required or included?
- How will it perform in challenging climatic conditions, including extreme heat or dust?
- How long is the battery life? Can you buy spare or additional batteries?
- Can you access maintenance and servicing support? How much will this cost?
- Have you ordered the appropriate probes? (External, adult and paediatric sized, not transoesophageal)
- Have you ordered other consumable items including ultrasound gel, red-dots for ECG?
- How physically robust is the machine/ travel case if it is to be used in remote settings accessible by bad roads, by boat or in dusty or humid conditions?

Specialised services

Advance radiology and medical equipment is sometimes used for monitoring and managing RHD. For example, cardiac catheterisation can also be used to assess valve function and cardiac pressure measurements in settings where it is available.[24] Similarly, cardiac magnetic resonance imaging (MRI) is used to gather more information about valve damage and heart function – however, this is not the routine standard of care in most endemic countries and the value of additional information remains under investigation.[24] Specialised equipment and laboratory services will be needed for visiting surgical teams or when establishing cardiac surgical facilities. Guidelines and minimum standards for equipment and laboratory support services for cardiac surgery exist for developed settings and may be adaptable to low resource settings.[153]

5. INTEGRATION AND HEALTH SYSTEMS

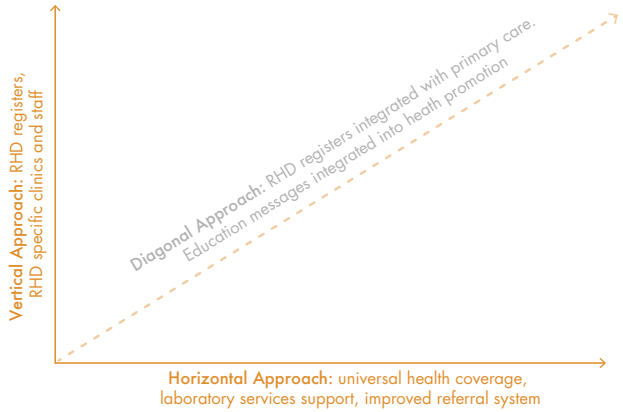
THINGS TO CONSIDER

- How do people at risk of – or living with – RHD interact with health services in your area?
- Does your programme have formal integration activities with other disease specific programmes?
- Do you consult with other groups and departments when planning activities, programmes and activities?
- Are primary care doctors and health workers supported to diagnose and manage RF and RHD?
- Are other clinicians easily able to contact your programme for advice or referral?

Introduction to integration

Integration of RF/RHD services into health programmes and services is widely recommended to prevent development of 'unsustainable monolithic programmes'.[22,154-156] Integrative models of care are thought to be important for sustainability, quality of care and accessibility for patients and communities. However, defining, describing and implementing integration in the context of RHD can be challenging.[156] In turn, this makes it difficult to develop an evidence base about the effectiveness of integration. Therefore, the theoretic benefits of integrative RHD care are yet to be demonstrated in empiric studies. This chapter outlines some of the rationale for integration of RHD control and some examples of attempts to deliver integrative programmes.

Figure 12: Models of Horizontal, Vertical and Diagonal Integration in RHD Control Programmes



WHO defines integration as 'the management and delivery of health services so that clients receive a continuum of preventive and curative services, according to their needs over time and across different levels of the health system.'[156] In practice, the term integration continues to mean different things to different people in different contexts, including:[156]

- Integration through **a package of services to particular groups** – defined by either age or disease category. Examples include Integrated Management of Childhood Illness (IMCI), Integrated Management of Pregnancy and Childbirth (IMPAC) or the Package of Essential Noncommunicable disease interventions (WHO PEN).
- Integration of clinical services **delivered from one geographic location.** In general this means ensuring that a wide range of services can be provided by a single local clinic. Functionally, this can be considered integration of services into comprehensive primary care.
- Integrative care may can also refer to **delivery of services over time**. This may reflect a life course approach to ensure that people receive care as their health needs change. For example, including antenatal care, delivery services and maternal care in a comprehensive programme.
- Integration is also used to refer to **embedding disease specific programmes within the broader healthcare system.** Disease specific services are generally called 'vertical' programmes. These differ from horizontal programmes which tackle more than one kind of disease through broad based programmes including primary care, laboratory services or medical record capacity building. The advantages and disadvantages of this approach are outline in Table 8.

Table 8: Merits of vertical and horizontal programmes (some content from[157])

	ADVANTAGES	DISADVANTAGES
VERTICAL	• Targeted, allows rapid results and 'easy wins'. • Outcomes relatively easily measured/quantified. • Health professionals can be trained to provide specialist services.	• Fragmented – people may need to visit many different clinics. Clinicians may not have complete information about each patient. • Potential for inefficiency and duplication. • May divert resources away from other diseases or consume all human resources. • Priorities may be influenced by international donors.
HORIZONTAL	• Integrative care reflects people's real needs. • More sustainable, less influenced by donor priorities.	• May be complicated to deliver and difficult to demonstrate outcomes.

In reality, a combination of horizontal and vertical programmes is usually needed; sometimes called a 'diagonal' approach.[158] Diagonal programmes strengthen primary healthcare and infrastructure alongside disease specific activities. For example, vertical HIV programmes have increasingly added capacity for other programmes in maternal health and non-communicable disease.[159] Control of RF/RHD is well suited to a diagonal approach.[160,161] Horizontal health systems components may include improved access to primary care, treatment of sore throats, access to laboratory services, referral pathways. Vertical components may include a register and dedicated staff to deliver secondary prophylaxis injections. This kind of diagonal approach may improve care for RHD while also supporting other broader goals in care delivery. For example there is some evidence that disease specific programmes can strengthen health systems if that is explicitly included as one of their goals.[162]

It may be helpful to think about the practical opportunities for integration in 2 two different directions:

- How can prevention or management of RHD can be usefully added to existing vertical and horizontal programmes?
- How can RHD programmes usefully provide or facilitate services which are delivered by other vertical and horizontal programmes?

Examples of this conceptual approach are outlined in Table 9.

These examples are explored in more detail across the rest of this chapter in Tables 10 and 11.

Table 9: A framework for describing integration of care delivery associated with RHD – adapted from Katzenellenbogen et al.[160]

	ADDING ELEMENTS OF RHD CARE TO EXISTING PROGRAMMES	ADDING ELEMENT OF NON-RHD CARE TO EXISTING RHD PROGRAMMES
Non-RHD vertical health care programmes, including HIV programmes, TB and NCD care.	• Considering RHD care as part of reproductive health services (for example, by screening women at high risk of RHD during pregnancy. • Adding RHD care to HIV programmes (for example, by facilitating delivery of secondary prophylaxis injections at HIV clinic appointments). • Adding RHD care to TB programmes (for example, by facilitating delivery of secondary prophylaxis injections alongside directly observed treatment, short course (DOTS).	• Providing dental care for people living with RHD to reduce the risk of endocarditis. • Facilitating screening of people living with RHD for infectious diseases, particularly ahead of surgical procedures. • Providing or facilitating access to contraception for people living with RHD.
Horizontal health services including comprehensive primary healthcare.	• Facilitating primary prevention (sore throat and skin sore management) through school-based care. • Increasing capacity of laboratory services to perform laboratory tests relevant to Strep A and RHD.	• Comprehensive primary healthcare for people living with RHD, potentially including: • Including risk assessment and management of NCDs in RHD clinical reviews. • Managing the complications of RHD including heart failure and arrhythmia.

Table 10: Adding RHD care to existing horizontal programmes

EXISTING VERTICAL PROGRAMMES	CLINICAL OPPORTUNITIES TO ADD OR INTEGRATE CARE FOR PEOPLE LIVING WITH RHD	OPPORTUNITIES FOR POLICY AND ADVOCACY INTEGRATION
ADDING RHD TO HIV PROGRAMMES Low resource communities with a high burden of HIV infection may also have a high burden of RHD – this is described in more detail in Chapter 21.	It may be possible to use existing infrastructure from HIV control programmes to provide RHD care. This has been best demonstrated in Uganda where physical resources from HIV facilities Joint Clinical Research Centre have been used to establish RHD register sites.[163] Similarly, in Kenya a pharmacy led anticoagulation service has a dual focus on people living with RHD and people living with HIV.[164]	Global efforts towards addressing HIV can be illustrative for RHD control given a similar distribution of disease burden and shared service delivery challenges. In particular, the experience of HIV highlights the importance of people living with a disease as advocates and in setting global disease control targets for RHD control.[165]
ADDING RHD CARE TO NCD CONTROL PROGRAMMES NCDs – defined by WHO as cardiovascular disease, diabetes, cancers and chronic respiratory diseases – caused 70% of deaths worldwide in 2015.[166] The growing number of deaths from NCDs prompted a High Level United Nations Meeting in 2011. This led to an international commitment to reduce premature mortality from NCDs by 25% by 2025.[167] Subsequently, all WHO Member States adopted the Global Action Plan on the Prevention and Control of NCDs (GAP).[168] Although the GAP focuses on NCDs caused by the four common risk factors for NCDs (tobacco, poor diet, inadequate physical activity and excessive use of alcohol), the GAP acknowledges the need to address RHD and other NCDs of childhood such as asthma, leukaemia and type II diabetes.[168]	Clinical addition of RHD services into NCD programmes generally focus on advanced heart valve disease. In particular, heart failure, AF and stroke, are common NCDs and complications are often managed through NCD clinics. For example, Rwanda has a strong focus on decentralising care to NCD programmes, including RHD care.[169] This approach means that 32% of patients being managed for heart failure through an integrated, NCD focused, echocardiography service had RHD.[170] In Kenya a novel programme to support NCD care by frontline health workers using mobile phone services included RHD.[171]	Some countries have included RHD in their national NCD Action plan – including Kenya, Rwanda and Pakistan[172-174] Secondary prevention for RHD is also included in the WHO Package of Essential Non-Communicable Disease Interventions for primary care in low resource settings (WHO-PEN).[175] Increasing access to cardiac surgery is also a shared goal in addressing ischemic heart disease and RHD.

ADDING RHD CARE TO REPRODUCTIVE, MATERNAL, NEWBORN AND CHILD HEALTH Women with RHD are at risk of heart failure and death during and immediately after pregnancy. Some women with RHD can not safely become pregnant and need reliable access to contraception. Other women will need close medical supervision throughout pregnancy. Some women with RHD will be diagnosed during pregnancy and there may be a role for screening women in early pregnancy to identify dangerous damage to the heart valves from RHD.	Increasing awareness of RHD and capacity for diagnosis/referral is a valuable addition to antenatal care programmes. Practical examples may include: • Provide education on RHD in pregnancy for frontline health workers. This may allow signs and symptoms of heart failure to be identified sooner during pregnancy and referrals initiated for specialist care. • Screening for a history of RF/RHD in routine antenatal care, including medical history and cardiac auscultation.[176] • Echocardiographic screening of pregnant women in RHD-endemic settings may be possible, usually timed around planned obstetric ultrasound. A pilot study of this approach has previously been conducted in Eritrea and a study is ongoing in Uganda.[176,177] The role of this kind of 'active case finding' in pregnant women remains the subject of ongoing research.	Policy and advocacy intersects between RHD and reproductive health include the importance of reliable contraception, early antenatal assessment and access to highly specialised delivery services.
ADDING RHD CARE AND SERVICES TO CONGENITAL HEART DISEASE PROGRAMMES Congenital heart disease (CHD) shares some similar issues with RHD: both affect young people, can be asymptomatic and lead to heart failure. Screening programmes for RHD (see Chapter 20) will inevitably detect a proportion of children who have CHD and some will require interventions.[178] Therefore clinical integration of childhood heart disease services and support services should encompass CHD and RHD.	Many of the same kinds of medications are used for RHD and CHD and cardiac surgery is a component of management for both conditions. In addition to medical needs the parallels between RHD and CHD also offer an opportunity for shared education and support for children and families. For example, in Australia, the charitable group Heart Kids provides camps, advocacy and support for families living with a range of childhood heart diseases.[179] In India, a joint programme for diagnosis and treatment of RHD and CHD has been delivered through schools.[180] In South Africa, RHD Action supported a 'Listen to My Heart' event including people living with both CHD and RHD.[181]	Opportunities for policy integration may include advocacy for access to paediatric cardiac services. New paediatric surgical programmes often begin by providing relatively simple surgical procedures to correct CHD lesions. Over time capacity for more complex surgery required for RHD can be delivered. CHD can provide an important advocacy angle for childhood heart disease because it occurs across the socioeconomic spectrum and may have particular impact for decision makers.

Table 11: Adding additional services to programmes provided for people living with, or at risk of, RHD

PROGRAMME TO BE ADDED TO RHD PROGRAMMES / SERVICES	OPPORTUNITIES FOR CLINICAL INTEGRATION
ADDITION OF REPRODUCTIVE, MATERNAL, NEWBORN AND CHILD HEALTH TO RHD CONTROL PROGRAMMES Women with RHD are at risk of complications during pregnancy. Therefore, supporting reproductive and antenatal health services is a valid consideration for RHD control programmes.	Practical examples of integrating reproductive care to RHD control programmes are emerging. Opportunities for integration include: • Educating all women with RHD that all pregnancies/deliveries need close medical supervision, and providing tangible support for accessing medical care.[182] One suggested model is adding family planning professionals to routine cardiac clinics.[183] • Developing a referral system for primary health workers and midwives to access echocardiography and specialist review for women with RHD. • In some places specialised cardiac-obstetric clinics provide shared care between cardiologists and obstetricians to manage high risk pregnancies.[184,185]
ADDITION OF DENTAL CARE TO RHD CONTROL PROGRAMMES RHD increases the risk of bacterial endocarditis, a serious infection of the heart valves. The bacteria that cause endocarditis are often found in the mouth and good dental hygiene is needed to keep teeth and gums healthy and to reduce the risk of bacterial endocarditis. This can be challenging for people living with RHD in low income settings.	Although there is a clear link between good dental care and RHD outcomes there have been few attempts to integrate these services. In New Zealand dental checks are provided at an integrated RF clinic.[186] In Kenya an outreach programme as part of the RF/RHD awareness and prevention project sometimes include dental services.[187] Other opportunities integration could include: • Including dental representatives on your advisory committee (see Chapter 2). • Partnering with a dentist working near your hospital or clinic to provide dental services • Included dental care on your priority-based care plan pathways (See Chapter 20).
ADDITION OF NCD RISK ASSESSMENT TO RHD CARE People with RHD are also at risk of NCDs, including ischaemic heart disease which can worsen heart function as discussed in Chapter 20.	RHD control initiatives may be able to embed NCD screening into routine care delivery – potentially alongside delivery of secondary prophylaxis, as part of priority based followed up (Chapter 19) or during preoperative evaluation surgical candidates (Chapter 24). Although older people with RHD often have coronary angiograms before heart valve surgery there are no published accounts of RHD programs providing NCD risk assessment as part of routine care delivery. Lessons about potential integration may be drawn from HIV programs and attempts to embed NCD risk assessment.[188,189]
ADDING HEALTH SKIN PROGRAMMES TO PRIMARY PREVENTION INITIATIVES. Although the association between Strep A skin infection and RF remains unclear (see Box 6) it is reasonable to consider including healthy skin programmes as part of comprehensive RHD control initiatives in some settings.	Clinical opportunities for addressing skin infections are most commonly packaged with primary prevention/sore throat initiatives. • In New Zealand some sore throat services (see Chapter 14) have expanded to deliver skin sore treatment programmes.[190] These programmes aim to tackle untreated skin infections, the commonest cause of medical hospitalisation in the high risk school age groups. This approach appears to have to provided good access to antibiotic therapy, facilitated referrals for other health issues and increased health literacy.[190] • Guidelines for management of skin infections may be added to training or education materials for sore throat treatment, taking swabs for culture or laboratory protocols.[191] Development of clinical guidelines for the management of skin infections in low resource settings is underway.[192] Clinical integration may be paralleled by advocacy, policy and research collaborations for the control of skin diseases.[193] For example, efforts to reduce household overcrowding are likely to reduce the burden of both skin infection and RF.

BOX 6:
What is the relationship between streptococcal skin infections and RHD?

Strep A pharyngitis causes RF and subsequent RHD.[194] Strep A also causes the skin infection impetigo (pyoderma). Observationally there is an overlap of communities with a high burden of impetigo and a large number of people living with RHD. In particular, Australian Aboriginal communities demonstrate a high incidence of RF – in these settings anecdotal reports of sore throat are generally low, Strep A throat carriage is very rare but impetigo is hyperendemic.[195,196] This correlation supports the idea that streptococcal skin infections, in addition to Strep A sore throat infections, may also cause RF or perhaps make individuals more likely to develop RF following Strep A pharyngitis.[197-199] There is some support for this population-level hypothesis in individual cases studies – a small number of individuals have developed RF following microbiologic confirmation of Strep A skin infection and in the absence of Strep A throat infection.[200] New data has also quantified the correlation between scabies infection, which often co-exists with Strep A infection, and the risk of RF or RHD diagnosis.[201]

Adding RHD to existing horizontal programmes which tackle a range of conditions

Adding RHD prevention and care to the health benefits package of universal health coverage

Universal health coverage (UHC) is 'ensuring that all people have access to needed promotive, preventive, curative and rehabilitative health services, of sufficient quality to be effective, while also ensuring that people do not suffer financial hardship when paying for these services'.[202] UHC has become a major focus of health reform and a WHO priority in recent years. In practice, implementing UHC often means defining a package of services that will be provided by the health system for all citizens – sometimes called a 'health benefits package'.[203] This process may provide an opportunity for primary, secondary and tertiary interventions for RHD to be included and to be funded in a way which makes them accessible to people in greatest need.[204] RHD Action has developed a detailed briefing document on 'Why RHD must be incorporated into universal health coverage' which can be downloaded online.[205]

Integrating prevention and care of RHD into routine primary care delivery

Primary care occurs at the 'front lines' of a health system – usually at the first place people go when they feel unwell. In RHD endemic settings, primary care is usually delivered in small local health clinics. Often these are staffed by community health care workers or nurses. They may be part of the public health system, non-government providers or be private clinics.

The foundation of primary healthcare is working at the community level, responding to a community's needs and taking into account the aspirations of each segment of a community at the economic, social and cultural levels.[206] The general principles of community-based programmes transcend disease specific issues and focus on the needs of individuals and their families.

RF and RHD are ideally suited to a primary care approach; early signs (sore throat, joint pain) are often identified by primary care clinicians.[207,208] Secondary prophylaxis and much ongoing care can be safely provided by primary care staff which reduces costs and improves accessibility. Providing the majority of care through supported primary care clinicians can benefit consumers and the broader health system. A number of specific control programme components can be delivered in the primary care setting:

- **Delivery of primary prophylaxis:** Evaluating and treating sore throats and skin sores is an important part of primary care and should be a core competency for front line health staff.
- **Identification of suspected RF:** Primary care staff have a critical role in identifying suspected cases of RF. Secondary prophylaxis can only be initiated for people who present for care and receive the correct diagnosis of RF. Diagnosis relies on accurate use of the Jones Criteria or local alternatives. In endemic settings primary care staff need sufficient training to recognise possible cases and refer for them definitive diagnosis.
- **Delivery of secondary prophylaxis:** Although register-based programmes are helpful for ensuring consistency of BPG administration delivery of the injections themselves is often a good fit with primary care clinics.[209]
- **Education and primordial prevention:** Primary care staff are uniquely positioned to know local families, identify who is at risk for RF and RHD, to provide education about overcrowding, advocate for families and provide targeted interventions where they are needed most. The important and time-consuming role of primary care in prevention, advocacy and education should not be overlooked amidst the delivery of clinical services.

Integration of education and prevention of RHD with school health programmes

School age children are at greatest risk of RF, making schools a valuable location for educating communities about RF/RHD. Teachers and educators have an important role in identifying children with a sore throat, symptoms of RF (particularly joint pain and chorea) and children with heart failure who are breathless and cannot keep up with their peers.

Prevention programmes, care delivery and specialist outreach may also be integrated into school programmes. Administrative or logistic support from schools to record secondary prophylaxis adherence, notify programmes of transfers or new students may also be possible.[211] Delivering education and services through schools generally requires support from the Ministry of Education, and often at an individual school level.[212]

A range of models for integrating RHD programmes with schools initiatives have been applied worldwide:

- In Cuba the education system was a key component of the plan developed to implement a control programme and included a representative of the Ministry of Education. Education personnel received training in RF and RHD.[33]
- In Zambia a workshop for 53 teachers was held to explore understanding of RHD and provide information about the disease. Of the participants, 45% had not heard of RHD prior to the event. Baseline awareness of the disease was low. Conversely, 24% reported knowing a school student living with RHD. Teachers participating in the workshop indicated a desire to contribute to RHD control initiatives.[213]
- School health nurses in South Africa are being trained to provide antibiotics to children with sore throats.[214] The role of the South African Integrated School Health Programme (ISHP) in supporting young people with chronic diseases, including RHD, is subject to ongoing discussion. Certainly the ISHP has a clear role in health promotion and awareness raising for sore throats, RF and RHD.[215]
- Diagnosis and management of sore throats and skin infections is integrated into schools in high risk communities in New Zealand as discussed in Chapter 14.
- A large school-based outreach programme in Kenya has been funded by the Mater Hospital since 2008. Large outreach camps include education, clinical service delivery, de-worming, support for secondary prophylaxis and screening for surgical services.[187]

> "The battle of RHD is not in the hospital. It is in schools and through treatment in health clinics."
>
> Dr. Marco Costa, Vice-President and Chief Innovation Officer at University Hospitals Health System speaking at the RHD Stakeholder Meeting, Kampala Uganda, October 2017.[210]

It is clear that integrative opportunities within school health exist. However, there are few examples of sustained school-based healthcare functioning at scale. For example, a School Health Programme survey in Tanzania alluded to the challenges of fragmentation and sporadic funding.[216] Pragmatic opportunities to amplify shared goals between school health and RHD should be identified and pursued where possible. Evaluation of these initiatives is an outstanding need.

6. GOVERNMENT ENGAGEMENT AND ADVOCACY

THINGS TO CONSIDER

- Does your programme provide a clear consistent message about RHD control priorities to local, regional and national governments?
- Do you have resources available to ensure that all advocacy activities are consistent and asking for the same outcomes?
- Can you provide high quality data that is relevant to the local population in a way that is understandable and usable by government bureaucrats and politicians?

Government policies underpin economic, development, housing, education and health outcomes in most countries. This means that governments have a significant impact on the socioeconomic conditions that people live in and which cause Strep A, RF and RHD.[217] Governments are also responsible for responding to Strep A, RF and RHD through health and other services. Therefore, government choices influence the cause of RHD and it's outcomes. For this reason engaging local, regional and national governments is an important element of comprehensive RHD control programmes.

Government agencies generally face a wide range of competing priorities and political imperatives. Government activities are further constrained by funding limitations, human resource capacity and bureaucracy. This is almost always an issue for health services in low-resource settings with a high burden of RHD and many other diseases to address. It can be difficult to ensure that RHD is recognised and control of the disease embedded within the health system. Authors of a recent review of RHD control opportunities note: 'Decision makers in these settings require up-to-date information about the epidemiology of Strep A, RF, and RHD as well as specific contextual information about local healthcare delivery patterns and barriers and facilitators to care.'[29]

This chapter provides an overview of strategies for engaging governments and other stakeholders to support goals or activities for RHD. This work is known as disease advocacy.

Planning advocacy

There are many different approaches to health advocacy.[218] One useful framework is structured around the following nine strategic questions and has been used by UNICEF.[219] This approach can be adapted for RHD specific use in a variety of contexts.

1. What do we want?

Effective advocacy requires a clear vision of what you are asking for and why you need it. This is sometimes called identifying an 'ask'. The ask will depend on your local situation, priority setting by your RF/RHD Advisory Committee and the stepwise conceptual framework outlined in Figure 4. Some of the preparatory work for fundraising (Chapter 3), including burden of disease estimates and a plan for intervention may also be helpful. The ask needs to be Specific, Measurable, Achievable, Relevant and Time-bound (SMART).

Specific advocacy requests for RHD control may include:

- Embedding Strep A, RF and RHD in national strategies such as the national NCD Action Plan.
- Allocation of funding or human resources, for example to provide someone to coordinate the RHD register.
- Permission to run training programmes or events.

2. Who can make it happen?

Identifying key decision makers will help to focus your efforts. Asking stakeholders about decision makers is a good way to get this information. The RHD Action Needs Assessment Tool includes a strategy for stakeholder identification, mapping and interview.[220,221] The approach outlined in the Needs Assessment Tool includes a systematic review, interview template and use of an online tool to help clarify relationships.

Different decision makers may be needed for different advocacy projects. For example, permission to include RHD training in schools may require input from the Ministry of Education. It is important to think beyond the health system when identifying who could help you achieve your goals.

3. What do they need to hear?

Clear, evidence-based messages underpin health advocacy. You will need to decide on how to frame your messages/evidence to have the greatest impact.

Many governments respond to data about the size of health issues. Delegates attending the 2016 All-Africa Workshop on RF and RHD identified the unmet need for data as a barrier to including RHD on the national health agenda.[99]

Some governments have responded to data on the costs of RHD which could be avoided with effective prevention strategies. For example:

- In Samoa in the mid 2000's overseas treatment for RHD surgery consumed up to 12% of the national health budget. This ongoing cost incentivised some government support for RHD control activities.[222,223]
- In Nepal the high costs of cardiac surgery helped the government decide to fund relatively low cost comprehensive control programmes.[224]
- In Uganda management of RHD accounted for half of the cost of treatment at the Uganda Heart Institute in 2015/16.[225]

Other governments have responded to evidence of inequality in the burden of RHD. For example, funding commitments in New Zealand reflected a political response to disproportionate disease burden in Māori and Pacific Islander people.[226]

4. Who do they need to hear it from?

Information about RHD and a focused ask can be presented by different kinds of messengers for different audiences. Some decision makers respond to evidence presented by technical experts, others are moved by personal accounts of the impact of RHD. Individual advocates are sometimes called 'RHD Champions'.

The role of RHD champions was highlighted in the World Heart Federation Position Statement on RF and discussed at subsequent meetings.[227] At the 3rd RHD Global Forum, an entire breakout session was dedicated to the role of RHD Champions: 'there was consensus across the discussion groups engaged in this topic that there are different types of roles and levels of champions and that within these roles are indeed a key 'spectrum of roles' ranging from clinical champions, political champions, and social champions who would appeal to a broad range of stakeholders engaged in or affected by RHD. It was noted that the key role of any champion is to raise the profile of the disease in multiple spheres.'[228]

Potential champions include:

- **People living with RHD:** The lived experience of RHD can have a tremendous impact on decision makers. Testimonials about the impact of disease are often more meaningful than facts and figures alone. People living with RHD around the world are becoming vocal advocates for the disease including in Fiji, Uganda and New Zealand.[229-231]
- **Community spokespeople:** In some settings the perceived health priorities of communities can influence government response and funding allocation. Demonstrating the concern of a community – and a commitment to disease control – provides a powerful signal of need. Petitions, calls to action or community consultation may provide additional opportunities for engagement.
- **Clinicians or researchers:** Clinicians can be powerful advocates for RHD control. For example, Professor Bongani Mayosi, a South African cardiologist and researcher has been a vocal advocate for RHD control.[232] Prof. Mayosi explains that large-scale studies proving disease burden were necessary to get ministers to throw their weight behind South Africa's Stop Rheumatic Fever campaign. 'A study in Soweto showed how big a problem this condition really is', says Mayosi. 'Ministers started listening, and the wheels of primary healthcare started to turn.'[232] In the Pacific Island of Tonga, Paediatrician Dr Toakase Fafakovikaetau has pioneered efforts to detect RHD early and establish a register based control programme with widespread support and multiple funding sources.[233] In Australia, Dr Bo Reményi's work in RHD research and advocacy saw her awarded the 2018 Northern Territory Australian of the Year.[234] This award provided opportunities for Dr Remenyi to advocate for RHD control to new audiences during media appearances and events.

Advocacy for RHD can be challenging, particularly for people with considerable clinical demands who may not have direct access to decisions makers. For example, one researcher in Australia reviewing a new RHD control programmes notes: 'In my discussions with clinicians, I came across some who had been championing this cause for years but despite their 'high interest', their 'low influence' meant that change was not so easy to make happen.'[235]

Visiting clinicians, particularly humanitarian cardiac surgery teams, also have an important opportunity to be advocates for specific goals. Visiting surgical teams have a rare and valuable opportunity to advocate for addressing the underlying causes of RHD.[236,237] To add sustainable value to their visits, surgical teams should be encouraged and supported to discuss the need for prophylaxis, pre-operative and post-operative care.[237]

- **Clinical organisations:** National clinical organisations have a credible professional voice to call for resources, attention and action to RHD. Policy or position statements can also be produced by professional groups, including medical colleges and associations, to advocate for a specific course of action. For example, the Australian Medical Association dedicated their 2016 Report Card on Indigenous Health to RHD and issued a 'call to action' to prevent new cases of RHD in Indigenous Australia by 2031.[238] In India the National Rheumatic Heart Consortium has been formed to advocate for national disease control priorities.[92]

5. How can we make sure they hear it?

There are many different avenues for communicating key messages. These may include face-to-face meetings, letters, events, campaigns and activities.

The 'stakeholder feedback' meetings conducted as part of the RHD Action Needs Assessment Tool can provide a good rationale for gathering decision makers together with clinicians and identifying next steps. The Uganda Heart Institute and RHD Action co-hosted a Stakeholders Meeting in late 2017.[239] The event included senior officials from the Ministry of Health, people living with RHD, clinicians, researchers, funding supporters and stakeholders from other countries in the region.

6. What do we have?

Consider the resources you already have for communicating your key messages – this may include photos, stories, data, graphs or powerful quotes. The RHD Action Resource Hub has a large number of briefing notes and information sheets – many of these can be adapted to reflect your local data or key messages.[240]

These resources may be suitable to combine into a fact sheet, briefing note, short video or other resource to share with decision makers.

7. What do we need?

You may identify gaps in information, resources, capacity or contacts which make advocacy difficult. Explicitly identifying these gaps and developing a strategy to address them may be helpful. For example, you may need to use your networks to secure an introduction to a key decision maker. Or you may need to arrange for high quality, consented photos of people living with RHD to illustrate their quotes and stories.

8. How do we begin to take action?

It can be helpful to focus efforts around a specific project or activity which can lead to bigger goals. This is one feature of the RHD Action Small Grants programme which resources small activities in the RHD Action Priorities Pyramid.[241] For example, RHD Action supported the 'empowering and supporting young people living with RHD activity' organised by the Fiji RHD Programme Team.[242] Thirty eight young people with RHD participated in the event which provided fun activities, education and a chance to meet other people living with RHD. Photos from the event can be used to continue illustrating the importance of RHD control so that young people with the disease can continue to live with minimal effects from the disease.

World Heart Day is a global initiative of the World Heart Federation and is celebrated on the 29th of September each year.[243] The day has a different theme each year but can be adapted to local priorities. RHD focused events provide an opportunity to tie activities with global goals in cardiovascular disease control. For example, in 2017 World Heart Day was marked in Cape Town by a symposium on heart disease in pregnancy and landmarks illuminated in red lights.

9. How do we tell if it is working?

Advocacy and government engagement rarely result in immediate success. It can take many years to build relationships, raise awareness and generate momentum. Changes in government, competing priorities and funding difficulties can all set back well planned advocacy activities. Often it feels like progress is moving very slowly. Nonetheless, small steps still move you forward. It can be helpful to review your advocacy plans and identify short and long term goals which can be revised or amended. Your RHD Advisory Committee may also periodically review Government engagement objectives.

BOX 7:

CASE STUDY **SUDAN**

Sudan is a lower-middle income country in Northern Africa with a population of 40 million people.[245] Sudan was part of the WHO Global Programme for RHD Control that was established in the 1980s.[246] Led locally by the Sudanese Ministry of Health,the programme focused on secondary prevention and supporting adherence to regular BPG injections. The first RHD control programme in Sudan concluded in 2000 and identified the following key lessons:[247]

- RHD control programmes need to be modified to involve primary as well as secondary prevention
- More advocacy is needed, namely by involving local and international non-government organisations, the public and the patients
- More internal will and cooperation with regional organisations from countries with similarly high RHD prevalence are needed in order to assure programme continuity

In 2012, the Sudan Heart Society established a new RHD control programme with the Sudan Ministry of Health.[4] The new programme focuses on improving primary and secondary prevention of RF and raising awareness of the condition. A National RHD Awareness Day is held on the 17th of July each year, involving events, educational materials, school-based programmes and media outreach.

Funding for various parts of the programme come from the WHO Sudan Country Office, Sudanese Children's Heart Society and the Sudanese American Medical Association.[249] The kind of relationship building and advocacy which can yield funding and technical support is exemplified by paediatric cardiologist Professor Sulafa Ali.

'Sudan's Ministry of Health was supportive of Ali's plan for an RHD programme but was surprised that she wasn't asking for funding initially. 'I felt at the start we just needed to get the project underway, and write and shout about it. The money would come later', Ali says. And indeed it did; once the programme was up and running, WHO's Sudan office funded the implementation, embedding it into a plan to tackle non-communicable diseases in Darfur.'[250]

A second edition of clinical guidelines for the management of Strep A, RF and RHD in Sudan have been developed.[247] A National Registry for RHD has been developed and initial patients enrolled from tertiary clinics with echocardiography facilities.[249,251] Over two years, 370 patients were added to the register, 82% of these people had severe RHD.[251] Secondary prophylaxis adherence remains an ongoing challenge – only 50% of people received 80% of their scheduled doses. Fewer than 10% of people who needed cardiac surgery were able to access it. Only 30% of patients received ongoing follow up care, reflecting difficulties in contacting patients and arranging ongoing review.[251]

The Sudan Programme has adopted a frame work of 'Surveillance, Integration, Collaboration, Advocacy & Training (SUR I CAAN)'. An echo screening project started in 2015, so far 12000 people have been screened using hand held echo machines, funded by research grants and charity organizations. An echo screening project started in 2015, so far 12000 subjects (children and adults) were screened using hand held echo machines, funded by research grants and charity organizations.[244] Using this data, RHD control sentinel sites are being established in Kordofan and Darfur. The programme was successfully integrated into the Ministry of Health Package of Essential Noncommunicable Disease (PEN) funded by WHO. Training modules were developed and used in PEN as well as other programs for physicians and medical assistants.

Regional advocacy

Groups of countries can act to tackle RHD collectively. This approach may facilitate sharing resources and expertise to address common challenges.

For example, a concerned group of cardiologists and cardiac surgeons from the Pan Africa Society of Cardiology committed to support RHD control efforts in a statement known as the 'Drakensberg Declaration' 2005, updated in 2011.[252]

In Pacific Island countries, clinicians developed a 'Call to Action' from a workshop held in Fiji in 2006, and again endorsed it at a subsequent workshop held in Fiji in 2008. The signed 'Call to Action' advocates five key messages to governments, international agencies, donors, non-government organisations and health care providers working in or with Pacific Island countries.[253]

Considerable regional leadership in RHD has emerged from the continent of Africa. In 2016 an All-Africa Workshop on RF and RHD was convened by the Pan-African Society of Cardiology and the African Union Commission. This event amplified consensus for addressing seven regional priorities in RHD control, subsequently adopted in the African Union resolution on RHD control.[99]

Global advocacy

Globally, a number of organisations are working to advocate for the RHD control agenda and provide technical support for implementing control programmes. More information about international efforts is available at rhdaction.org.

Additional resources

Further tools to help guide your advocacy and government engagement can be found online including the World Heart Federation Cardiovascular Disease and Development Advocacy Toolkit and the UNICEF Guide to influencing decisions that improve children's lives.

Table Mountain lights up red for World Heart Day 2016. Credit: @KMiljof / Twitter.

7. DISEASE NOTIFICATION

THINGS TO CONSIDER

- Are there notifiable diseases in your setting?
- Are there surveillance systems in place in your setting for notifiable diseases?
- Are RF or RHD notifiable?
- Are there other mechanisms in place in your setting that could capture cases of RF or RHD, if these conditions are not made notifiable?
- How do you define suspected or confirmed cases?
- If notifiable, can notifications be automatically added to the RHD register? (see Chapter 7)

A notifiable disease is any disease required by law to be notified to the government or other health authority. Diseases to be notified to WHO are outlined in the International Health Regulations but most countries have their own list of nationally notifiable diseases and frameworks.[254] For example, the WHO Regional Office for Africa has supported the implementation of Integrated Disease Surveillance and Response Strategy (IDSR) to monitor, prevent and control priority notifiable infectious diseases.[255] Making a disease legally 'notifiable' by doctors and health professionals allows for interventions to control the spread of highly infectious diseases such as influenza, poliomyelitis or yellow fever. In addition to infection control and prevention, notification provides a legal framework to collect information about the burden and distribution of disease, which allows for public health action.

Different countries use different criteria to decide whether a disease should be made notifiable. Often this reflects the size and severity of the disease.[256]For example, in Australia, there are four broad principles for assessing suitability as a notifiable disease.[257] Operationally, these are further refined into detailed criteria.[258]

Reviewing these principles shows that RF meets the broad criteria for suitability as a notifiable disease in endemic settings (Table 12):[257]

RF has been made notifiable in many places: New Zealand, [259]most of Australia,[44] South Africa, [260]and Fiji.[261] RF was notifiable in the USA from 1956 to 1994.[262] RHD is also a notifiable disease in some parts of Australia[263] South Africa (from 1989 to 1992).[260] Including RHD as a notifiable condition provides a more comprehensive epidemiologic picture. It also provides a mechanism for strengthening care for people who have ongoing care needs (through secondary prophylaxis) but in whom the first diagnosis of RF may have been missed.[263]

Table 12: Screening criteria for suitability as a notifiable disease[257]

A recognisable disease	RF can be diagnosed by the modified Jones criteria. A different case definition may be used to trigger notifications, particularly in settings where full application of the Jones Criteria is prohibitively difficult.
A preventable disease	High quality secondary prophylaxis can alter progression to RHD following an episode of RF. Primary prevention with antibiotics can prevent the first episode of RF.
There must be the potential for action	Action can be taken at all levels for RF and RHD – primordial, primary, secondary and tertiary. For individuals with RF, notification can be linked to a register-based programme (see Chapter 16) which provides secondary prophylaxis and prevents recurrences of RF.
There must be an identified population or sub-population target	Young people at greatest risk of RF often come from vulnerable communities and/or Indigenous populations.
Notified data should be usable	RF notifications offer an opportunity to understand the distribution and burden of disease, plan interventions and monitor outcomes.

Case definitions

Many infectious notifiable conditions can be identified from positive laboratory tests (direct laboratory notification). There is no blood test for RF or RHD, so cases must be diagnosed and notified by clinicians. A strict case definition and accurate clinical diagnosis are needed to make this possible. However, clinicians often have considerable demands on their time and may be unfamiliar with reporting requirements. These barriers mean that underreporting is common in many settings.[260,264]

Clear guidelines are important for communicating and disseminating case definitions and pathways for reporting. The New Zealand 'Communicable Diseases Control Manual' is a good example of this approach – the RF chapter includes: case definition, case classification, notification procedure, case management and contact management.[259] If RF is going to be notifiable in your setting the threshold for reporting will need to reflect local diagnostic standards and access to diagnostic resources (including blood tests, ECG and echocardiography).

Notification systems

Wherever possible, if RF is going to be notifiable, it should be integrated into existing disease surveillance systems and cross referenced with the RHD register. Some factors to consider are whether any additional staff or resources are needed, whether any changes will need to be made to reporting systems and who else needs to be involved to streamline this process. Multiple reporting pathways can cause confusion and fragment fragile systems.[264] For example, in South Africa, suspected cases of RF are notified to the National Institute for Communicable Diseases on paper-based forms or through online notifications.[265]

Closing the feedback loop and engaging with stakeholders

It is important to engage with clinicians and other agencies/stakeholders of the notification system early so that you have their support from the very beginning. One of the barriers to clinicians reporting RF or RHD can be a perception that data gathered is not used to make changes.[261,264] This can cause frustration, particularly if clinicians are being asked to spend time collecting epidemiologic data.[264] Reporting information and action back to clinicians may be helpful to demonstrate that reports are being collated and acted upon. Routine publication of notifiable disease surveillance is standard in some parts of the world, for example via the Pacific Public Health Surveillance Network. Your programme may be able to provide updates on the number of cases of RF identified through the programme.

Opportunities for integration

In countries without strong notifiable disease programmes, clinicians with an interest in RF/RHD may work with other potentially notifiable diseases to develop or strengthen reporting systems.

8. HUMAN RESOURCES

THINGS TO CONSIDER

- Do you have an 'RHD Person' to coordinate your programme?
- What priority tasks will your programme address? Who can complete these tasks?
- How will members of the RHD team communicate with each other?
- What are the major areas of workforce strength and shortage in your area?

Human resources for health are limited worldwide. There is an estimated shortfall of 18 million health workers needed to achieve the Sustainable Development Goals.[266] This absolute shortage is amplified by the relative shortages in developing countries with the highest burden of RHD. The World Heart Federation Roadmap for RHD control identifies human resources as one of the eight roadblocks for tackling this disease.[101]

Comprehensive health workforce planning is outside the scope of TIPs. The World Health Organization has an extensive set of resources and tools to support health workforce planning in a range of settings. These can be accessed online at www.who.int/hrh/en/

This chapter offers some guidance about tasks and potential roles within comprehensive RHD control. Resources are often limited and it is rarely possible to employ an 'ideal' set of staff. It may be more useful to think about the tasks that need to be completed, and then identify people who can be responsible for different components.

Individual nurses and health workers are commonly identified as the most important elements of successful programme delivery, as outlined in Box 7. Wherever possible, these key individuals should be supported by a number of clinical and non-clinical staff, as exemplified in Box 8.[267]

BOX 8:

The importance of having an 'RHD person'

Descriptions of RHD control programmes over the last 60 years have revealed the importance of a single key contact for programme implementation. Sometimes this person is called the programme manager, the nurse manager or the register coordinator. Irrespective of title, having a single core person dedicated to developing and delivering the programme is a key component of care delivery, continuity and medication adherence.[268-270]

"As a nurse in Uganda, I spend most of my time caring for patients affected with chronic cardiovascular illnesses such as congenital heart disease, myocardial infarction, and rheumatic heart disease (RHD). My typical day starts at 7am and ends at 9pm. During this time, my work involves updating the RHD registry with new patients, those that have died and identifying those that are lost to follow-up. I then spend the day in the outpatient clinic counselling patients, enrolling patients in the RHD registry, and administering Benzathine Penicillin injections in the Coumadin [anticoagulation] clinic which I run concurrently."

Samalie Kitooleko, Nurse In-Charge of Uganda Rheumatic Heart Disease Registry, 2017[271]

Supporting people working in critical RHD coordination roles is important for retention, programme sustainability and institutional knowledge.

"The programme manager can liaise effectively with schools, school health services, primary healthcare centres and maternal and child health services, as well as with departments of medicine and paediatrics and laboratory services in hospitals."

Dodu and Bothing, Rheumatic fever and rheumatic heart disease in developing countries, 1989[272]

BOX 9:
Case study – Kiribati[273]

Having a dedicated RHD coordinator in the Pacific Island of Kiribati has demonstrated how effective this role can be especially in the early stages of an RHD programme. Within eight months of commencing the Kiribati RHD programme and employing a dedicated RHD nurse coordinator, first year screening was conducted, national protocols were finalised, over 170 RHD cases were identified and added to the new RHD database, 154 health workers were trained, community awareness campaigns were conducted and education materials developed in local language and disseminated. Patient injection cards were distributed, Benzathine Penicillin injections books were provided to all clinics and standing orders were introduced to RHD patients to reduce their wait and travel times each month. Similar results are evident in other countries in the Pacific region that have employed a dedicated coordinator including Fiji and Samoa. Although good progress has been made over the same time period in other countries, particularly in Tuvalu and Nauru, there is a notable difference in what can be achieved with a dedicated RHD coordinator/nurse.

Specific human resources

Community health workers

Community health workers have different names in different countries. In general they are defined by WHO as 'individuals who should be members of the communities where they work, should be selected by the communities, should be answerable to the communities for their activities, should be supported by the health system but not necessarily a part of its organisation, and have shorter training than professional workers.'[274] In some places, community health workers are more commonly referred to as Frontline Health Workers (FLHW) – though frontline staff can include some professional cadres.[275]

Community health workers underpin service delivery in many regions where RHD is endemic. Community health worker programmes have proven effective in addressing specific health issues in a range of low resource settings.[276] The skills and community connections of community health workers are also well suited to delivering elements of comprehensive RHD control programmes. For example, in some part of Australia, Aboriginal Health Practitioners deliver secondary prophylaxis injections and support follow up plans for people living with RHD.

In one programme in India, health workers have been trained to consider the diagnosis for RF and support referrals to other services.[277] Developing models of community health worker care sore throat treatment, identification of RF and management of RHD is a global priority in RHD control.[278] Sharing stories, best practice and resources for community health worker care will help to amplify and accelerate the role of health workers.

Nurses

Health systems across the globe are faced with a critical shortage of nursing staff. Furthermore, the distribution of nurses means they are often not available in areas of greatest need. The causes of nursing shortages and maldistribution are multifactorial. Wages, education and training and access to medication and essential medical equipment are all areas of priority in rectifying the global nurse shortage.[279]

Nurses can assume critical roles in RHD control programmes, including management of service delivery and clinical leadership. For example, framework for a senior nurse practitioner role has been developed in Australia.[280] Nurses acting in independent roles need support for their practice. For example, in a nurse-led primary prevention RHD control programme in New Zealand a manual of operations is used by all providers and standing orders are in place for the registered nurses for treatment of defined conditions by a delegated authority.[281] Similarly, in Haiti, nurses lead an anticoagulation clinic for people who have had valve surgery for RHD. Using clear protocols nurses are able to deliver high quality care and safe anticoagulation.[282]

Midwives

There are not enough midwives to deliver maternal care in low resource settings – a 2014 report identified major deficiencies in midwifery care in 73 countries.[283] Where midwives are accessible they can play an important role in identifying symptomatic RHD in pregnancy and referring women for specialist assessment. For example, in Australia training has supported midwives to 'think RHD' in the 'at risk' population, and trigger referrals for cardiac echo's to diagnose RHD.[284] Ensuring that midwives have access to training about RHD and are able to refer for further assessment is an important opportunity for integrative care delivery.

BOX 10:
Traditional healers

Traditional, faith-based or community healers are a significant part of healthcare in many parts of the world. Including traditional healers has proven to be very important to the customs of some groups of people. In some cultures, the healer is a respected elder, part of many families and someone with a gift that is to be respected.

People with Strep A infections, RF and RHD from around the world report seeking traditional health care. For example:

- In Samoa nearly 10% of people identified the village healer as their first intervention for sore throat.[285]
- In Cameroon 45% of people attending hospital with heart failure (predominantly from RHD) had already consulted a traditional healer.[286]
- Traditional beliefs have also impacted management of RF or RHD in Hawaii,[77] Nigeria,[287] Zambia[288] and Rwanda[52] and are likely to be influential in many other settings.

Traditional therapy is frequently perceived as delaying diagnosis and treatment. Delays in the treatment of sore throat and diagnosis of RF may compromise outcomes of clinical care. Understanding the role of traditional healers in your setting and the opportunities for education, partnership or referral may be an important determinant of programme outcome. It may be possible to include traditional healers in your education programme or advocacy activities.

Doctors and medical specialists

The global shortage of specialist clinicians is particularly acute in areas where RHD is endemic.[289] Clear, consistent messages about the need for specialty staff – and the impact of limited human resources – help to keep these issues on the national and international agenda. For example, in Rwanda there are only 2 paediatric cardiologists for a population of 10 million people.[290] In South Africa, there is a shortage of at least 2000 fully qualified cardiologists to meet population needs – in addition to existing issues with maldistribution of the workforce.[291] To achieve a target of one paediatric cardiologist for every 500,000 people, a further 20 paediatric cardiologists would be needed in South Africa.[291]

Similarly, cardiothoracic surgeons are rarely available in low and middle income settings. A suggested ratio of cardiothoracic surgeons is 1 per 800,000 population. In South Africa, the current ratio is one surgeon per 4.5 million people.[291] Recent estimates suggest there are only 135 cardiothoracic surgeons in 14 countries working across sub-Saharan Africa.[292] The small absolute numbers of surgical providers are amplified in paediatric populations.

Sustainable funding to operate training programmes, funding for support personnel, such as medical and nursing staff is difficult in environments where finances are limited. The demand is high and some have suggested that only large scale political and socioeconomic change will see developing nations realise the change that provides greater access to paediatric cardiothoracic surgery.[293]

Echocardiographers

Skilled echocardiographers (sonographers) with significant training and experience in cardiac views can be a very valuable addition to well-developed RHD control programmes. Good echocardiography services can free up time for cardiologists and assist with triaging people for intervention. There is very little information about the global echocardiography workforce; partly because training programmes and definitions are difficult.[294] Assessment and accreditation of echocardiography services can be an important step towards standardising and supporting this essential part of the health workforce.[295]

Strategies for retaining staff

The global health workforce shortage can make it difficult to retain health staff[296] – particularly when staff have been trained or have special skills. Your programme may be able to work with staff and develop a retention strategy allowing for addressing training, promotions and conditions which make it more likely that key individuals will continue in the programme. See Table 13 for an overview of factors influencing health worker recruitment and retention.

"Retaining health workers can be done by providing adequate salary, something as simple as clean water, key supplies and working equipment for patient care, clear job descriptions and supervision and at least some form of continuing education."

LeBlanc, Creating a global climate for pediatric cardiac care, 2009.[297]

RHD and other programmes in developed settings should be aware of the WHO Global Code of Practice on the International Recruitment of Health Personnel.[299] This provides guidance about minimising 'brain drain' from developing settings through responsible recruitment practices.

Table 13: Factors contribution to health worker recruitment and retention.[298]
These factors contribute to health worker migration n by encouraging departure (push) or encouraging recruitment to a new setting (pull).

REMUNERATION	Differential between source and destination country can be a significant motivator to leave and barrier to return.
EMPLOYMENT OPPORTUNITIES	Availability of jobs and job security during times of budget cuts and public service retrenchment.
PROFESSIONAL DEVELOPMENT	Migrating for improved access to opportunities that will progress a clinician's career or training. Home settings do not have employment opportunities to use newly acquired skills.
WORKING ENVIRONMENT	Excessive workloads, poor working conditions, low staffing levels and human resource systems that are inadequate for the environment.
RECRUITMENT STRATEGIES	Shortages in destination jurisdictions have triggered migration rules to be changed to allow for strategies to recruit from other nations.

Strategies for supporting and sharing clinical roles

Telehealth

Telehealth generally refers to activities which provide clinical support, connect users who are not in the same physical location, involve different kinds of information technology and share the goal of improving health outcomes.[300] Although telehealth capacity is often constrained in resource limited settings with a high burden of RHD some research and pilot programmes have demonstrated proof of concept. In Egypt, the Aswan Heart Centre uses telehealth consultations with primary care clinics for initial consults prior to people travelling for clinical assessment.[301] Telehealth has also been a critical component of scaling up access to echocardiography screening programmes for RHD by allowing remote review of echocardiograms.[302] The role of telehealth in echocardiography screening programmes is discussed further in Chapter 20.

Telehealth may also facilitate international consultations and training of clinicians in low resource and/or geographically remote settings. For example, the Pacific Island Health Care Project has been supporting services for some Pacific Islands since the early 1980s.[303] One telehealth programme between Angola and Portugal has supported cardiac clinical decision making and triage for surgical services.[304] Similarly, in Malawi a partnership between the Kamuzu Central Hospital and Texas Children's Hospital provides telehealth training and clinical consult with international colleagues.[305] This partnership has also spurred efforts to support the Malawi Ministry of Health to implement a register-based RHD control programme. In Uganda, a donated telemedicine facility make it possible to share echocardiography images and consult with team partners in the United States.[145]

Local policies and governance of telehealth projects can be complex and often require detailed consultation. This is an evolving area of clinical practice with changing norms and expectations. Some contemporary guidance is available on these issues – including a statement on 'Telemedicine in Pediatric Cardiology' from the American Heart Association.[306] Programmes considering telehealth activities should review evolving guidelines and seek technical support for implementation.

Task shifting and task sharing

Task shifting – sometimes called task sharing – is commonly identified as a way of addressing human resource shortages in health. In general, task shifting is a process of delegation whereby tasks are moved, where appropriate, to less specialised health workers. There is some evidence that this approach can be effective and affordable for non-communicable disease programmes in developing countries if coupled with health system restructuring.[307] Growing evidence suggests that task shifting in low resource settings can also be cost effective and efficient.[308]

In RHD the major focus of task shifting has been to train new, non-expert, operators to undertake limited view screening echocardiography for early diagnosis of RHD.[309,310] The role of non-expert operators for echocardiography screening is discussed further in Chapter 19. There may also be a role for upskilling non-cardiology clinicians (in general practice and obstetrics) to use focused echocardiography techniques to triage and refer people living with RHD.[291]

However, there are many other opportunities for nurses, community health workers, clinical officers and other frontline health workers to share tasks and improve service delivery and RHD control goals. Task sharing can include delegating authority to frontline health workers to prescribe medication, dispense/administer medication, initiate referrals or manage follow up.[27] Collating and evaluating case studies of these practices will be important for identifying best practice for task sharing in RHD.[311]

Safe and sustainable task shifting requires careful planning and support for all staff. Often, clinical protocols are needed to define which tasks are being delegated. Regulatory change may also be needed to ensure that less trained staff have legal protection for undertaking activities which may be outside their historic scope of practice.[307] For example, in Brazil only physicians are allowed to perform echocardiography and make a diagnosis.[312] The opportunities for task shifting and sharing are significant if well planned, well resourced and coordinated with the needs of local health services.

9. HEALTH WORKER TRAINING

THINGS TO CONSIDER

- Which health workers need to know about RF and RHD in your area?
- What do they already know, what kind of training have they received?
- How many people do you need to train?
- Do they already have planned meetings that you could incorporate training into?
- Are there universities, post-graduate training providers or specialist training programmes which could amplify your message?
- Are there novel opportunities to include remote, telehealth or online approaches to education and training materials?
- How can training material be evaluated and improved?

Informed and engaged healthcare workers are a critical component of successful disease control programmes. Baseline awareness of RF/RHD varies in different settings:

- In Kenya in 1989 a survey of 55 enrolled nurses and clinical officers indicated a very limited understanding of Strep A sore throat or RF.[313]
- In Tanzania in 2011, awareness of 540 primary healthcare workers was good with high levels of insight into appropriate treatment for Strep A sore throat, clinical presentation of RF and recommended duration of treatment.[314]
- In Sudan in 2015, surveys of doctors found that knowledge about sore throat, RF and RHD was average - with an average score of 50% on a quiz about the topics.[315]

Without training of healthcare workers diagnosis may be missed, clinical guidelines will not be used effectively and quality of care delivery is likely to be variable. However, providing training can be difficult in settings with many competing health priorities and service delivery demands. Provision of training may also be expensive and consume considerable resources without necessarily delivering expected results.[316] This chapter focuses on strategies to support training for frontline healthcare workers in primary care with a focus on RF and RHD control.

Develop an education and training plan

RHD control programmes should support all health staff to improve knowledge, expertise and skills in the prevention, diagnosis and management of Strep A infections, RF and RHD. Education, training and the dissemination of information increase capacity and improve outcomes.[317]

An education and training plan is helpful for prioritising training needs, allocating resources and evaluating impact. The plan should include an assessment of training needs, objectives, target staff for education, expected competencies and outcomes.[318] For example, 'The Primary Health Care Package for South Africa – a set of norms and standards' defines the following expectations for primary healthcare staff:[319]

- Suspect streptococcal infection of the throat following a complaint of acute sore throat with the finding of pharyngeal exudate and tender cervical glands.
- Suspect and refer acute rheumatic fever by recognition of polyarthritis, heart murmur, arthralgia, fever, erythema marginatum, chorea, subcutaneous nodule, history of sore throat in last month or previous rheumatic heart disease.
- Recognise and refer possible rheumatic disease by murmurs and previous history.
- After definitive diagnosis in hospital and notification ensure patient receives prophylactic treatment.

Baseline needs assessment for training can be extracted from healthcare provider interviews conducted as part of the RHD Action Needs Assessment Tool. This interview tool includes questions about current levels of training and some clinical scenarios to identify knowledge gaps.

Planning considerations following needs assessment may include:

- The number of staff who require training.
- The baseline education, numeracy and literacy of those staff.
- Barriers and challenges faced by those staff.
- Time and facilities available for training.
- Resources and teaching materials which may be needed.
- People available to provide training information.

Training should incorporate knowledge assessments before and after information is provided. Participants should also have the opportunity to provide feedback on elements they found most useful or enjoyable. This makes it possible to assess whether education programmes are achieving their goals and to refine future events.

Opportunities to deliver training for frontline health workers

Embed into existing training courses

Ensuring that RF and RHD are included in materials for existing local community health worker, nursing, midwifery and medical training is an integrated and relatively low cost intervention. For example, in Australia, consultation with midwives identified RHD as a gap in existing midwifery curricula. Training resources were developed and disseminated to address this gap.[320]

There may be a delay between instituting training and new graduates entering the workforce. Providing access to education and training for clinicians and health workers already working in high risk settings is a valuable way to improve diagnosis and management.

Training courses

Courses dedicated to the diagnosis and management of RF and RHD provide a focused approach to share knowledge. They have been very successful in some settings for improving clinical management.[321,322] A training manual for an RF/RHD workshop in the Pacific Islands has been developed, this resource provides a sample timetable, teaching and evaluation resources. In Nepal, 1500 health workers, including those joining the secondary prophylaxis programme, received training on the management of RF/RHD.[323] Training of doctors in Sudan demonstrated that, following a series of lectures, there was a marked increase in knowledge about Strep A treatment, identification of RF and treatment of RHD.[315]

Clinical skills for management of RF/RHD may also be needed in some settings. Skills may be best taught in person, in focused events, potentially with simulation of clinical events and with real-time feedback. For example, training on safely delivering antibiotic injections for prevention of RF and management of anaphylaxis has been an important part of RHD control efforts in Zambia.[324] In Australia, RHD training courses include clinical and practical case studies about a range of scenarios in RHD management.[325]

However, bringing people together, especially for RHD training, can be expensive and may interrupt the provision of health care in settings where human resources are limited. One approach to this problem is a train-the-trainer model. This allows a smaller number of staff to receive content knowledge alongside skills for training other people – allowing for a 'cascade' of information to spread.[326] A train-the-trainer approach to RHD has been developed in Nepal with twenty-six people completing the programme as of 2016.[327] The training manual for this programme is available online.[323] Alternatively, training formats which allow clinical staff to learn at their own pace and at convenient times may be needed.[328]

"I think there is a definitely a need to educate doctors about ARF/RHD, especially in the Northern Territory. This is particularly important to staff who are moving here from other states or countries, who may not have come across these conditions before. The stakes are so high when the condition is misdiagnosed or inappropriately managed and for those reasons alone education is essential. I felt quite unprepared to diagnose and manage ARF/RHD when I initially moved to the Northern Territory and I hope the transition for future staff can be made more comfortable."

Dr Thilini Basnayake, participant in the Northern Territory Australia Rheumatic Heart Disease Education Workshop, 2016.[329]

Training modules

As part of the Public Health Training Initiative in Ethiopia, formal training modules have been developed to educate healthcare professionals at all levels about RF/RHD. Core modules complement satellite modules for specific professional groups – public health officers, nurses, laboratory technicians, environmental health technicians and community health workers.

Similarly, in Australia, a suite of online modules have been developed for clinicians to improve their skills in particular areas: dental, echocardiography diagnosis, medical management of RHD, primary and primordial prevention, anticoagulation, RHD and pregnancy, screening for RHD, secondary prevention and valvuloplasty. Although the modules focus on the Australian setting they are accessible to users around the world (once you have registered for access). Online modules are also used to train New Zealand health staff about rheumatic fever but are not accessible beyond the health department.[330]

Online training modules about RF/RHD have been also developed for international settings by WiRED, with a focus on community health workers and school teachers.

Education and awareness raising poster for healthcare workers. This *Think ARF* campaign was developed by the Queensland RHD Register and the RHD Control Program.
Credit: RHD Programme Queensland / Queensland Health.

Publications

Publications, mail outs, posters or other resources can be used to share information for health professionals. Posters can provide an opportunity for people at risk of RF and RHD to engage with clinicians and facilitate conversations about the disease. For example, Mrs Phyllis Tokarua from Fiji recounts her experience taking her child for clinical review. Her son was subsequently diagnosed with RF, facilitating delivery of secondary prophylaxis.

"My son was in my arms unable to walk because of the pain in his joints. I started to look at a poster on the wall. I could see the arrows pointing to different joints of the child in the poster. The doctor later asked me 'what's wrong with your son' and I said I think he has that (at the same time pointing at the poster in the nearby waiting area)."

Mrs Phyllis Tokarua from Fiji, 2018

Similarly, in Zambia posters describing the symptoms of Strep A throat infections are used to remind health staff of features which require treatment.[331]

It may also be possible to involve frontline healthcare workers in community education about RF/RHD. This means that healthcare workers gain knowledge on key topics while becoming familiar to the local community, which may support health seeking behaviour.

Posters and other training information can be downloaded from the RHD Action Resource Hub.

"A sustained information and educational effort to professionals and residents has been at the core of the Program. Methods used have included the media, professional journals and mailings to physicians. For more than two decades of the Program's existence 1317 physicians have availed themselves of the Primary Prevention Services offered by the city."

Levinson et al, Chicago Rheumatic Fever Program, United States of America, 1982.[332]

Training health workers in settings with a high prevalence of RF and RHD involves initial extensive consultation and familiarisation with the health workers themselves, knowledge of training opportunities already available, and an understanding of the context within which the health workers are working. If possible, making use of training structures already in place and embedding RHD into the local curriculum is useful, as well as utilising online and overseas training courses already developed for this purpose. Promoting community awareness, as outlined in Chapter 11, is crucial for encouraging health-seeking behaviour.

BOX 11:
The difficulty of high staff turnover

Communities with a high burden of RF and RHD may be in areas with a high turnover of health workers.[333] This makes ongoing training – and the development of accessible resources – particularly important. Training and material should cover essential clinical knowledge and the mechanism of how your RHD control programme works.

In particular, new staff should be made aware of the symptoms of RF and the need for specialist evaluation to confirm diagnosis. Every episode of RF that goes unrecognised is a missed opportunity to begin life changing secondary prophylaxis.

Education on how to notify cases, referrals and interventions may also be needed. Orientation for new staff is particularly important if they have come from high resource settings where RF and RHD is rare.

Opportunities to access training for specialist clinicians

In places with specialist services additional training may be needed for specific clinical skills. It can be difficult to facilitate training of doctors and specialists alongside delivery of clinical services. Sometimes a period of exchange or international placement is needed, although this can disrupt provision of local care. For this reason, the Pan-African Society of Cardiology has developed a system of modular cardiology training focusing on different clinical components: cardiac pacing, echocardiography and interventional cardiology. These can be undertaken at different times and at different institutions that offer the PASCAR curriculum.[102] Online specialist training modules focusing on heart valve disease are also available, although these are focused on developed settings. In-person training opportunities include the African Paediatric Fellowship Programme which provides networked opportunities for specialty and subspecialty training, including paediatric cardiology, throughout the African continent.[334] Other specialist training occurs alongside visiting teams and international collaborations, often in conjunction with cardiac surgery services. These programmes are addressed in more details in Chapter 25, Provision of International Services.

Opportunities for integration

Including RF and RHD into local clinical protocols and handbooks provides a comprehensive orientation for new staff and a teaching programme.

10. PROGRAMME EVALUATION

THINGS TO CONSIDER

- Do you have a system for monitoring or evaluation of your programme?
- Do you have clearly defined, realistic goals or outcome indicators?
- What kind of reporting requirements do you have to donors, government or other groups?
- Do you seek feedback from your patients, clients, communities or people living with RHD?
- Do you have a budget for programme evaluation?

Monitoring and evaluating your RHD control programme is critical for:

- Understanding whether your work is having the desired impact.
- Identifying areas which need to be revised or improved to better meet the needs of your community.
- Setting or revising targets.
- Reporting to donors or funding agencies.
- Reporting to communities and people living with RHD.
- Improving clinical outcomes.

Comprehensive RHD Control Programme Evaluation

Planning monitoring and evaluation should begin when you begin planning your RHD control programme. Planning for evaluation should incorporate input from your Advisory Group about the activities, objective and goals which are most important and should be routinely measured.

Monitoring and evaluation is a specialised discipline and expert advice may be needed to inform the monitoring strategy for your RHD control programme. Resources outlining the process and best practice of evaluation may be helpful, including the United Nations Development Programme (UNDP) 'Handbook on planning, monitoring and evaluating for development results'.[336]

RHD Action has used the principles of this UNDP resource to develop a framework for monitoring and evaluation of RHD control programmes as part of the Needs Assessment Tool.[28] This approach makes it possible to use baseline data collected during the needs assessment process to identify performance-based measures for process improvement. The NAT recommends that 'Baseline data should be compared against measurable outcome target goals (both quantitative and qualitative) that determined by consensus of the stakeholders'.[336] Detailed information and sample data collection tools are available to be downloaded online.

Inviting or commissioning external review of your RHD control programme can provide an important independent perspective on progress towards agreed goals. Evaluation findings may also be useful for reporting to funding agencies or government stakeholders on RHD programme activities and performance.

Narrative reviews have identified a number of valuable lessons in South Africa, Kenya and the Top End of Australia.[337,338] However, narrative reports are usually structured as free text which can make it difficult to compare quantitative measures of progress between sites or over time. Detailed evaluations have been commissioned in high income settings including Australia and New Zealand[40,96,281], and in the Pacific.[339] This kind of professional evaluation usually costs money and it may be necessary to reserve some programme budget for evaluation activities.

Table 14 : Defining monitoring and evaluation[335]

MONITORING	EVALUATION
MONITORING involves continuous checking of the programme to ensure that it is proceeding according to plan. Monitoring is conducted by collecting data (indicators) at regular intervals on: • what programme activities are being undertaken (process indicators). • the extent to which programme objectives are being met (outcome indicators). • progress towards the programme goal (impact indicators).	EVALUATION 'provides an independent and in-depth assessment of what worked and what did not work, and why this was the case'.[336]

Other forms of monitoring

Clinical audit

Clinical audit is 'a quality improvement process that seeks to improve patient care and outcomes through systematic review of care against explicit criteria and the implementation of change'.[340] Clinical audits in low resource settings are a valuable opportunity to improve quality of care, though it can be difficult to initiate and act on audit outcomes in some settings.[341]

Audits are used to monitor quality of care, and can be useful for evaluating how well your RHD programme is delivering planned services. Often they involve review of clinical records or register data, and compare the care delivered against recommended standards. Audits have been completed to assess service delivery or adherence to guidelines in a number of RHD control programmes.[342,343] Surgical outcomes require a specialised data base approach outlined in Chapter 24.

Continuous quality improvement

Improving the quality of healthcare delivery is an essential consideration for low resource settings. Continuous quality improvement (CQI) is an action research process that has been implemented in many industries, including engineering and manufacturing. It shares similarities with the clinical audit process, though CQI tends to be more comprehensive and designed to be an ongoing project. CQI in health includes implementing systems of care based on best practice guidelines, researching the level of adherence to the guidelines, and reflecting upon the results. The process is cyclical and incremental, therefore measures to improve practice can be implemented and evaluated, and through their participation in the process, team members integral to improvement increase their knowledge.[344]

An extensive package of CQI interventions for RHD programmes has been developed in the Northern Territory of Australia.[345] Protocols for clinical audit to inform CQI in this setting can be downloaded online. In this setting 'CQI also provides a structure to refine and reinvigorate programmes to promote sustainability'.[346] Therefore CQI may provide an opportunity to introduce the idea of evaluation into existing programmes in way which is participatory and not confrontational.

Although there are not yet a set of internationally agreed audit indicators, the Key Performance Indicators (KPIs) developed by RHD Australia provide an example of potential CQI benchmarks.[347] Your programme will need to select indicators relevant to your setting. It may be most practical to identify a number of representative sentinel sites where indicators can be monitored more closely.[317]

BOX 12:

Ensuring experiences of people living with RHD are captured during evaluation

Evaluation should include the views and experiences of people receiving services from the programme. Qualitative, semi-structured interviews have been most commonly used to explore satisfaction with health services.[348] As many people living with RHD are young and some are from vulnerable communities appropriate ways to obtain their views should be utilised – potentially including interviews, focus groups, drawings and other creative options.

Ethical considerations in monitoring and evaluation

There is a range of ethical issues that should be considered in the design and implementation of monitoring and evaluation activities, such as avoiding conflicts of interest, privacy and confidentiality, and transparency. While public health programme monitoring and evaluation activities are often not subject to the independent ethical review process that is undertaken for research, it is nonetheless critical that such issues be considered in designing and implementing these activities.

An important example of this is the collection of data from those working with, and accessing, the health programme. Interviews or focus group discussions may be undertaken with health workers and sometimes with patients. These perspectives can be very useful to understand what aspects of a health programme are working well, and what components could be improved. It is critical that the information and perspectives contributed are treated confidentially, and that there be no perceived or actual impact on respondents from their participation and provision of honest feedback. Such considerations must be addressed in the design and conduct of an evaluation, and also in decision-making around who will have access to the data collected, and how the findings will be used and disseminated.

An ethical framework for monitoring and evaluation of public health programmes has been developed, which may be a useful guide for how to address these issues in developing your RHD programme monitoring and evaluation activities.[349]

PRIMARY PREVENTION

Primary prevention encompasses interventions to prevent the development of RF. Typically, this has entailed treatment of Strep A infections in young people. Prompt treatment of Strep A sore throat with effective antibiotics can prevent the development of almost all cases of RF. Although some cases of RF appear to occur without young people recalling a recent episode of sore throat, the opportunity to prevent RF, and preclude development of RHD offers promise for disease control.

Delivery of antibiotic primary prevention requires attention to a number of biomedical and systems challenges. Evaluation and treatment of sore throats requires that families seek medical care, that appropriate antibiotics are prescribed, and that antibiotics are taken as directed. In highly endemic settings, families and health systems face many competing demands on time and financial resources. Sore throat may be considered a benign childhood illness which is too mild or too frequent to warrant medical care. Community education is an important way of ensuring that families are aware of the risk of RF from untreated sore throat, and to provide information about accessing the appropriate health services. Management of sore throat is an important role for primary care and community health services. Guidelines are needed to support these health professionals to evaluate sore throats, and to provide appropriate treatment when indicated. Although a single dose of injectable antibiotic (BPG) is highly effective, some guidelines provide for an oral treatment option of 10 days duration. Adherence to twice daily antibiotic tablets complicates delivery of effective treatment to prevent RF. RHD programmes have an important role to address each of these issues, and to bring families and health workers together to tackle sore throats. Programmes should identify and address barriers to primary prevention; this may include support for community education, clinical guidelines, access to appropriate antibiotics and strengthened primary care services. In some places, barriers to primary prophylaxis have been addressed by incorporating some health care delivery into schools, including the diagnosis and evaluation of sore throats.

Development of a Strep A vaccine has the potential to revolutionise primary prevention by preventing Strep A infection and subsequent development of RF. A vigorous research agenda to develop a Strep A vaccine has persisted over a number of decades, and has yielded some signs of promise. Sustained investment, clear demand and a strategic framework for vaccine development is needed to support development of a market-ready vaccine. Although the technical components of vaccine development are outside the remit of most RHD control programmes, the RHD community should not be passive participants in the vaccine agenda. Control programmes have a vital role in collecting epidemiologic data, articulating the unmet need for a Strep A vaccine, and advocating for ongoing research and development. Few other groups bear witness to the ongoing human toll of RHD or can so effectively advocate for population level interventions. As research continues, RHD control programmes will be important stakeholders in consultation, to ensure vaccine candidates are acceptable and accessible to communities in need.

Primary prevention is the prevention of RF after Strep A infection. Currently, this means providing appropriate antibiotics to children and young people with Strep A infection. Prompt antibiotic treatment of sore throats can prevent the development of almost all cases of RF following Strep A.[350]

Primary prevention is very effective in reducing the risk of RF in individuals, however there are many barriers to primary prevention at a population level. These are well illustrated in the Continuum of Care, developed by the Medtronic Foundation , which underpins the Needs Assessment Tool developed by RHD Action.[28]

Continuum of Care For RF and RHD

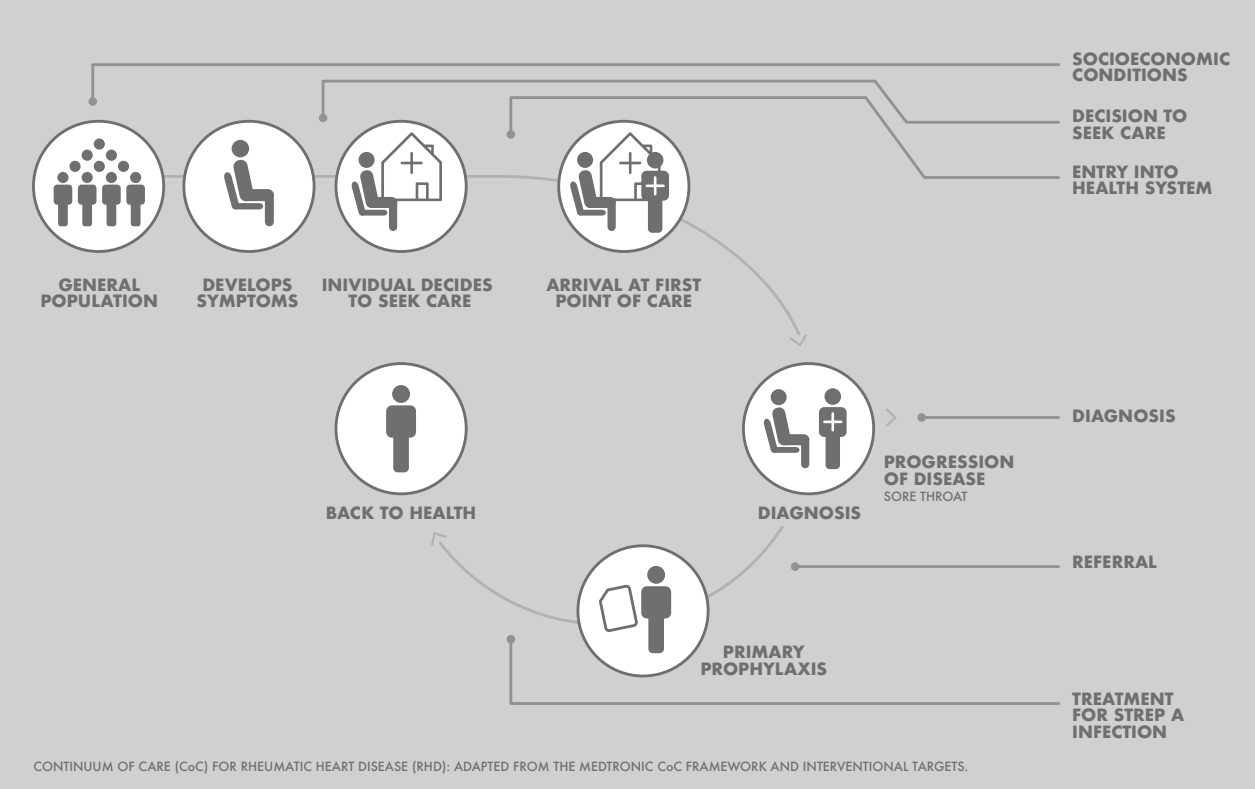


Figure 13: Continuum of care for sore throat

Delivery of antibiotics requires a decision to seek care, access to healthcare, health worker knowledge of the causes of sore throats, appropriate choices about antibiotic use, access to those antibiotics and that antibiotics are taken as directed.

This chapter reviews some of those key challenges and suggested approaches. In particular, Chapter 11 outlines approaches to community education, given that awareness of RF and RHD is a major contributor to the decision to seek health care. Chapter 14 outlines opportunities and experience with active case finding for Strep A infections.

"South Africa has the conditions that are required for success in eradicating RF and ending RHD. The National Department of Health has prioritised the prevention of the disease through initiatives such as National Rheumatic Fever Week, the inclusion of RF among notifiable conditions, and efforts to improve access to primary healthcare. Health practitioners need to play their role by notifying cases of RF, treating all children with a sore throat with penicillin according to the South Africa guidelines, and entering all our patients with RHD in the electronic register".

Mayosi, National rheumatic fever week: the status of rheumatic heart disease in South Africa, 2016.[351]

11. COMMUNITY EDUCATION AND ENGAGEMENT

THINGS TO CONSIDER

- Who needs to learn more about sore throat, RF and RHD in your community?
- What key messages do they need to know?
- How will you test messaging to ensure they make sense to the target audience?
- Who can help you develop education and engagement materials?
- How will you disseminate the education materials to the target audience?
- How will you evaluate the impact and reach of your communication?
- How will you sustain your communication with target groups over time?

Organisations, communities, families and individuals are critical stakeholders in the control of RF and RHD. Primary prevention of RHD requires an engaged community that knows when and how to seek healthcare. This can be difficult because the relationship between Strep A to RF and RHD is complicated. Unlike other diseases – for example, malaria, tuberculosis or respiratory infections - the association between cause (sore throat) and outcome (heart damage) is often poorly understood.[352]

Linking sore throats to joint pains of RF and subsequent RHD is a critical component of community education. There is no single best way to develop an education programme; this chapter aims to provide some options for you to consider in the context of local needs and preferences.

Considerations when developing an awareness campaign

WHO has identified a number of components of successful community health awareness campaigns.[353] These components are outlined below with reference to RF/RHD specific education.

Participant involvement

Community members and organisations should be involved in the design and implementation of education programmes. This helps to ensure that health messages are valuable, culturally appropriate and best reflects the local needs.[352] For example, you need to be sure that people can read or interpret your message.[285] Community consultation, focus groups or piloting of materials are important ways of engaging the community. Community representatives on your Advisory Committee (see Chapter 2) may be also be a valuable source of feedback.

Planning

Careful planning is required to develop a successful community education strategy. A clear and memorable message should be identified early, a target audience defined, methods of message dissemination and the goals of the campaign established. Clear messages reduce confusion and improve retention.[353] Single sentences or phrases can be helpful, for example: 'Sore throats can lead to a broken heart', 'stop sore throats hurting heart' or 'fight the fever' in New Zealand.[354,355]

Needs and resources assessment

Existing community resources and infrastructure should be identified and evaluated to avoid duplication. New resources may be developed to meet the needs of your target audience: School students, teachers, families, communities, healthcare professionals, government officials can all be the focus of an information campaign. Each population will require a different approach, reflecting their existing knowledge and information required.

Comprehensive programme

Comprehensive RHD control programmes address community education at a variety of levels; including children, parents, teachers, students and community groups. Consistent messages should be developed which can be tailored to the needs of each specific target audience without contradiction or confusion. The epicentre for RF/RHD prevention should not be solely in the health clinic, but at schools, homes, faith-based centres and community spaces.

Integrated programme

Integrated education programmes provide consistent messages across all media and locations. For example, billboards can be used to reinforce key messages from radio advertisements. In this way, health messages can be integrated into people's lives.

Long-term change

A successful programme will establish a sturdy foundation that can serve as a platform for enduring change. Community education about RHD should be established as policy to promote sustainability, rather than occur as a single outreach effort.

Research and evaluation

Education campaigns should be evaluated during and after their implementation, to improve existing awareness programmes and inform the design of future campaigns. Pre and post intervention surveys can demonstrate acquisition of new knowledge.[285]

Opportunities for integration

Partnering with communication experts

Developing health promotion material and ensuring it provides a useful message to the right people is communication science. Campaigns can be expensive and poorly considered messaging can cause unexpected problems or unintended consequences. Partnering with communication experts can be a valuable way to minimise risks and maximise outcomes. You may be able to access support through hospitals, health departments, businesses, universities or charitable groups.

Materials and media options for RF awareness

RHD Action hosts a **Resource Hub** online. This hub is a collection of existing RHD materials categorised by format, intended audience and language. The resources are freely downloadable and reusable. The collection is constantly being added to as new material becomes available. If you develop resources for your programme please consider sharing them with RHD Action so they can be further used and adapted.

Pamphlets

Pamphlets and printed handout are relatively low cost, easy to distribute and can be taken home by families for later reference. However, experience from New Zealand suggests that few families (only 35%) had read pamphlets about RF taken home from school.[356] You may need to develop brochures in a range of languages or images suitable for low literacy settings.

Posters, billboards and paintings

Billboards, sidings, posters and public notices may be a valuable – and relatively low cost - opportunity to provide health promotion messages.[285,357] For example, the Kenyan Heart Foundation ran a programme to paint 'Talking Walls' in schools displaying signs and symptoms of RF.[338] Billboards have been widely used by Nepalese RHD control programme.[327]

Film and video

Videos and films can be a powerful way of sharing information about RF and RHD. A number of educational resources have been developed for use in schools, community meetings and TV advertisements. In 1996 an RF/RHD information video was produced for the Northern Territory of Australia, funded by charitable donations.[113]The video was widely distributed to community health centres. When it was evaluated some years later 90% of nurses or managers reported the video to be a valuable educational tool.[113] Contemporary film projects include the Take Heart movie providing information and insight about the lived experience of RHD.[358]

Producing videos can be time consuming and expensive, so it is important to have a clear vision for how these materials will be used and where they can be distributed. Interviewing or involving policy makers in the film may be a way to engage their interest in RHD control. You can find existing video resources about RHD via the RHD Action Resource Hub or through the RHD Action YouTube Channel.

Online or technology based

Electronic community education modules may provide an important new medium to engage groups at risk of RF and RHD. An interactive digital teaching module on RHD was used in Kenya. Animated presentations linked sore throat to RF and then RHD. When the programme was evaluated it was found that school children participating in the module had statistically significantly more knowledge about RHD than their peers, and retained this knowledge after one week.[359]

Social media

In some parts of the world Facebook, Twitter and similar services are very popular with young people at risk of RF and RHD. Social media campaigns which encourage sore throat treatment, share knowledge about symptoms of RF and RHD and raise awareness about the problem have the potential for significant impact.[352] There is an active online community of organisations and individuals sharing information and events about RHD. On Twitter this includes @RHDAction and@RhEACHout. On Facebook the RHD Action Facebook group provides information focused for people living with RHD.

Radio and television

Radio messaging may be particularly useful for dispersed populations or in times of social instability and in areas of low literacy. Radio messages about RF and RHD has been used in New Zealand,[360] South Africa,[361] Nepal, [357] and the French Caribbean.[34] Programme evaluation from New Zealand found radio messages had been well retained by the target audience. Radio stories from New Zealand can be downloaded online.[362] Radio items to raise awareness of RF could include:

- Interview with doctors or visiting specialists.
- Perspectives of people living with RHD.
- Discussion with celebrities.
- Advertisements about RF, RHD and sore throat management.

Media training for doctors or others who are going to be interviewed on radio and television can be helpful to support clear communication of key messages. In Darfur, Sudan, a programme to raise awareness of RHD also included training for journalists on technical specifics of the disease.[363]

Newspapers, magazines and print media

Starting scrapbooks or folders of media coverage about RHD can be a useful way of ensuring media messages are clear, consistent and regular. Articles may be useful for evaluation and tracking trends in media coverage.

Performances

In some education programmes children have been encouraged to develop songs, skits and poems focused on RHD.[364] In Tanzania, a song 'MoyoWetu' has been composed to raise awareness of RHD among primary school children.[365]

Celebrity engagement

Celebrities can provide a powerful message about the importance of RHD control.[290] People who are famous or popular may be able to access adolescents who can be difficult to reach with traditional health promotion messages. In New Zealand, sporting heroes living with RHD have been powerful disease advocates.[366] In Fiji, a contestant in the national 'Hibiscus Pageant' shared her experiences living with RHD and has used her profile to spearhead an awareness campaign about RF /RHD.[367]

Events

A wide range of events are possible to increase awareness of RF and RHD. In South Africa the first week of August has been marked as 'Rheumatic Fever Week' for over 25 years.[368] This week provides a focus for media and health promotion activities. In Sudan, a National RHD Awareness Day was marked with the distribution of posters and pamphlets in 2015.[369]

Christine Katusiime, Vice-Chair of the RHD Support Group Kampala, Uganda speaks at a 'Listen to My Heart' event with other people living with RHD.

Education for Specific Audiences

School students and teachers

School-based education is an effective strategy to target young people most at risk of RF and RHD.
Education can be incorporated into regular activities:

- Health education classes.
- School books or notebooks.
- Peer education programmes.

In the New Zealand setting presentations by health care professionals were more effective at communicating with students than either paper handouts or lessons from a primary school teacher.[370]However, engaging teachers with the programme appears to be important for the transmission of knowledge from teachers to students.[370]

In Sudan, teachers receive a manual about RHD and hundreds of teachers have seen a video about RHD as part of their training.[371] Part of Cuba's successful RF/RHD control programme was school-based education of pupils, teachers and parents on the importance of prevention, symptom recognition and adherence with secondary prophylaxis.[33] In Nepal, students have written poems, performed skits and sung songs about RF/RHD to raise awareness.[357]In Samoa, visiting teams provide a detailed education package to school children and families alongside local doctors.[372] This Samoa package included a puppet show, RHD-related lyrics set to familiar tunes and a coloring book with information for parents on the final page. In Kenya, school student exposure to a training module about RHD appeared to increase awareness on post-exposure testing.[359]

The Nepalese RHD control programme has included an RHD educator in programme staff.[327] This RHD educator is a teacher trained in RHD who educates children, teachers and communities about RHD. Programmes can have one or more RHD educators. However, this strategy is limited by school attendance – in settings where few children attend or complete school this approach may fail to deliver education to those in greatest need.

Integrating health messages into school curriculums can be a sustainable way of ensuring that children receive consistent health messages each year. Materials and activities should be planned and designed in collaboration with education officials to ensure that they can be integrated into their programmes.

Young Professionals Chronic Disease Network (YPCDN) volunteers conducting awareness raising activities about RF and RHD in Uasin-Gishu County, Kenya as part of their RHD Action Small Grant Programme.

Opportunities for integration

Health education for young people at risk of RF and RHD should include a comprehensive package of health messaging. This may include a range of health and hygiene messages; hand hygiene, healthy eating and tobacco control education.[352] Health messaging can occur alongside other education initiatives. In Kenya, for example, school children were taught about RF/RHD in conjunction with healthy diet education.[364]

12. SORE THROAT DIAGNOSIS AND TREATMENT GUIDELINES

THINGS TO CONSIDER

- Are there standardised guidelines for diagnostic protocols and clinical management of sore throat in your area?
- Are children and families in your setting likely to be adherent with a full 10 day course of oral antibiotics?
- How could adherence with oral medication be supported or improved?

Sore throat (pharyngitis) is a common childhood disease in most countries. In settings with a high burden of RF/RHD approximately two thirds or more of these sore throats are caused by viral infections. Viral infections do not require antibiotic treatment. However, in children, up to third of sore throats are caused by a bacterial infection, most commonly Strep A.[373,374] In adults only about 10% of sore throat infections are caused by Strep A. Adults are also at a lower risk of RF. Therefore primary prevention is usually focused on young people, usually school aged, or defined in some settings as aged 3–35 years.[375]

Antibiotic treatment of Strep A throat infections in young people can dramatically reduce the risk of RF. Specifically, treatment with oral penicillin can reduce the attack rate of RF following Strep A by about 70% and up to 80% with an intramuscular injection of penicillin.[350] A full 10 day course of appropriate oral antibiotic treatment, or a single dose of intramuscular BPG injection, started within nine days of sore throat symptoms can prevent almost all cases of RF. [350,376,377]

In settings where RF and RHD are still common, diagnosis and antibiotic treatment of streptococcal pharyngitis are critical. However, in developed countries where the risk of RF is low antibiotics are generally not indicated.[378] Deciding who should receive antibiotics is challenging because it is not possible to reliably distinguish viral sore throat from bacterial sore throat on clinical history and examination alone. It is not feasible or desirable to treat all children with sore throat with antibiotics - overuse of antibiotics increases the risk of side effects, antibiotic resistance and inefficiency in resource allocation.[379] Although, in some settings with high rates of RF/RHD, guidelines do recommend that all children who present with a sore throat should receive antibiotic treatment; a strategy that preferences the importance of ARF prevention over the negative effects of antibiotic over-treatment. Hence, a preferable strategy would be one that distinguishes which children are most likely to have Strep A infection causing sore throat and therefore have the best chance of benefiting from treatment.

A number of diagnosis and treatment guidelines for pharyngitis have been published by organisations in high resource settings.[12,378] Although these provide useful resources, recommendations vary between settings and are not necessarily directly applicable to low and middle income countries with a high burden of RF.[380] In addition, the clinical presentation of sore throats varies significantly between low income settings.[381]

Developing local guidelines for the diagnosis and treatment of sore throats is an important role for RF/RHD control programmes and is discussed in this chapter.

Why develop (or adapt) local clinical guidelines for Strep A infections?

Establishing local guidelines is an important way to:

- Standardise treatment and reduce decision making demand on health staff.
- Rationalise the use of antibiotics to minimise the risk of adverse drug events and antibiotic resistance.
- Ensure that communities receive consistent messages about when to seek treatment.
- Strengthen ownership of the guidelines and improve use by clinicians.
- Facilitate task shifting and task sharing in some settings (see Chapter 8, human resources).
- Deliver care which meets the needs of local communities.

Different programmes and places will make different choices about Strep A treatment guidelines. These tend to reflect local opinion, experience and resources.[382] Local guidelines usually try to strike a balance between identifying the greatest proportion of Strep A infection, minimising costs of diagnostic tests, minimising unwarranted use of antibiotics.[16]

Acute rheumatic fever and rheumatic heart disease: Sudan's Guidelines for diagnosis, management and prevention.

It may be possible to embed local guidelines into existing standardised treatment protocols. For example, WHO has identified 'management of sore throat' as possible adaptation to the Integrated Management of Childhood Illness (IMCI tool).[383] Inclusion of sore throat in IMCI has occurred in Turkey, a number of Pacific Islands and Vietnam.[384,385]

Some RHD-endemic, low resource settings have developed local guidelines for Strep A infections and RHD management. For example, in Sudan the Federal Ministry of Health, Sudan Heart Society-Working Group on Pediatric Cardiology, Sudanese Association of Pediatricians and Sudanese Children's Heart Society have collaborated to develop the second edition of national guidelines for the disease.[386]

In New Zealand local sore throat guidelines have been developed and are augmented by a flow chart to summarise detailed reference material for clinical use.[375]

Primary prevention guidelines need to address two main areas: method of diagnosis and method of treatment. These issues are outline in brief below.

Diagnosis of Strep A pharyngitis

Broadly, there are four main approaches to deciding whether sore throat symptoms in children in RF endemic settings are likely to be caused by Strep A outlined in Table 14.

Table 14: Strategies for diagnosis of Strep A throat infection

CLINICAL ASSESSMENT	Clinical review then treat	In some settings, brief clinical review is used to determine whether children have features of a viral infection such as a runny nose or a cough. For example, in South Africa, children with reddened tonsils and fever receive antibiotics provided they do not also have a runny nose or hoarse voice.[387] A large scale approach to primary prevention in Costa Rica also used short clinical review as the criteria for treatment.[35]
	Clinical scoring tool then treat	A range of clinical scoring tools have been developed to try and distinguish Strep A pharyngitis from viral pharyngitis.[388,389] These appear to have some use in identifying which sore throats are most likely to be caused by Strep A. However, the most widely used scoring tools tend to be highly sensitive (correctly identifying children with Strep A pharyngitis) but poorly specific (incorrectly identifying children with viral infection as having Strep A pharyngitis). This means that many children will receive unnecessary antibiotics.

TESTING In a test-and-treat model, patients with a sore throat have antibiotic therapy guided by biologic tests to distinguish Strep A pharyngitis from viral pharyngitis. This approach supports rational use of antibiotics but may miss some cases of Strep A sore throat. It is also limited by availability, cost, feasibility and other practical issues, particularly in low and middle-income countries.	Culture for Strep A and treat	The best (gold standard) laboratory test for whether a child has Strep A pharyngitis is a throat culture. To conduct this test the clinician rubs a sterile swab across the tonsils and pharynx of a symptomatic child. The swab is sent to the laboratory and used to grow bacteria from the back of the throat. The presence of Strep A in the laboratory sample indicates that Strep A was present in the child's throat and was the likely cause of infection. [See Box 14 for further details about carriage]. The main disadvantages of this approach are the cost delay to access results. Growth of the bacteria in the laboratory takes at least 48 hours. This makes culture impractical for making day-of-review clinical decisions. In some places antibiotics are provided to children who have had a throat culture taken - if the culture result is negative the caregivers can be contacted by phone and advised to stop taking the antibiotics as they are no longer needed. The accuracy of throat swabbing is variable and depends on swabbing technique, transfer times and laboratory practices.[135,373] Accuracy of throat swabs is also reduced if the person with sore throat has taken antibiotic treatment prior to swab collection. This can be problematic in places where antibiotics are readily available without prescription. These variables are important considerations if throat swabs are going to be part of your RHD control programme and expert advice may be needed to manage laboratory systems and logistics.
	Rapid strep test for Strep A and treat positives	'Rapid tests' for Strep A have been developed to allow for faster diagnosis than with classical laboratory culture. These rapid tests work by detecting parts (antigens) of the Strep A bacteria and are sometimes called rapid antigen detection tests (RADT). Results are available at point of care, usually within 5–10 minutes. Collection of the sample is the same procedure as pharyngeal swabs for culture. A 2016 Cochrane systematic review found that relative to throat culture, RADT tests in children with sore throat have a sensitivity of 86% and specificity of 95%.[373] This means that the RADTs give a false negative result in 14% of cases and children may miss out on antibiotics they would benefit from. RADTs give a false positive result in 5% of cases and children may receive antibiotics they do not need. However, the performance of individual RADTs was highly variable by test used and collection technique. In general, use of RADTs seems to improve rational use of antibiotics - they may reduce unnecessary dispensing and ensure Strep A positive children are most likely to receive antibiotics. These changes may improve outcomes and reduce costs in some settings.[390] The costs of RADT vary between settings, though direct consumable costs are usually higher than for throat culture. Cost-effectiveness is likely to depend on the cost of antibiotic therapy, alternative diagnostic approaches, incidence of RF and performance of the test itself. In addition, some guidelines recommend a confirmatory throat swab to be completed as well as a RADT test – further increasing costs.[373] The evidence for RADT continues to evolve as new technologies improve the available tests. There is little global experience with wide scale use of RADT as part of population-level primary prevention programme. Pilot studies to date have been disappointing.[391] Programmes developing local guidelines for primary prevention should review up-to-date published literature and seek specialist advice.

BOX 13:
Antibiotics resistance and primary prevention

Doctors are sometimes worried that treating too many sore throats with antibiotics will cause antibiotic resistance.[350,392] Use of penicillin to treat sore throats has not been associated with penicillin resistance in Strep A; no Strep A isolate has ever demonstrated penicillin resistance. The mechanism of this persistent susceptibility to penicillin is poorly understood but has been maintained for many decades with widespread use.[393] There is the potential for overuse of antibiotics for pharyngitis to contribute to resistance in other bacteria. This risk is increased if penicillin or amoxicillin are replaced by broader-spectrum antibiotics. For example, Streptococcus pneumoniae is a major cause of pneumonia which has demonstrated resistance to penicillin following widespread overuse of antibiotics to treat for viral infection.[394] It is important that systems to support rational prescribing and accurate diagnosis of Strep A are in place and updated regularly. The use of broad spectrum antibiotics for pharyngitis should be discouraged.

BOX 14:
Colonisation and carriage with Strep A

A small number of children have Strep A bacteria detectable in their pharynx without symptoms of active infection or evidence of immune response on blood tests.[395,396] These children are describe as having 'Strep A carriage'. There is no evidence that children with persistent Strep A carriage are at risk of RF provided they remain asymptomatic.[397] It is possible that children with persistent Strep A pharyngeal carriage could be infectious to close contacts, although overall carriers are thought to be substantially less contagious than actively infected children.[397]

The mechanism by which some children have Strep A present in their throat without active infection is somewhat unclear.[395,397] The proportion of children with Strep A carriage varies in different communities and with age. Overall, in a meta-analysis of asymptomatic children in a variety of settings 12% were found to have Strep A carriage.[374]

Strep A carriage causes clinical uncertainty when there is a concurrent, but unrelated, viral throat infection. In these cases, children will have a positive throat culture or RADT but are unlikely to benefit from antibiotic therapy because Strep A is not really the causative organism. Different programmes respond to this clinical dilemma in different ways.[397]

Management of Strep A sore throat

Penicillin remains the first line antibiotic for the prevention of RF following Strep A sore throat in most settings.[12,46,375] Penicillin can be administered orally as penicillin V or as an intramuscular injection as BPG. Adherence is also improved with one-time BPG injection because strict adherence to a twice-daily oral regimen for 10 days is difficult to maintain for many patients.[398] Once-daily amoxicillin is an alternative to twice-daily penicillin V, although its efficacy in RF primary prevention has not been proven. Erythromycin is suggested as an alternative for patients with a proven history of hypersensitivity reaction to penicillin.[12,46,375]

Appropriate pain relief and supportive care should also be addressed in sore throat guidelines.[375]

If your guidelines include oral antibiotic options it may be worthwhile to consider how individuals and families can be supported to complete the full course of medication. Strategies may include:

- Provide a clear verbal explanation to continue antibiotics, even if symptoms resolve.[190]
- Provide memory cues or visual aids for each day of tablets. For example, New Zealand children taking oral primary prophylaxis are provided with a fridge magnet with 10 days marked and a set of stickers to put on the magnet each day as antibiotics were taken.

BOX 15: Guidelines for treatment of Strep A skin infection

There is some evidence for an association between Strep A skin infections and RF (see Box 6).Therefore, treatment of skin infections is sometimes included in primary prevention guidelines including the successful French Caribbean programme.[34] Guidelines for skin infection management can be included in IMCI and other standardised protocols to enhance care.[400]

Table 15: Appropriate antibiotics treatment of Strep A pharyngitis

	BENEFITS	DISADVANTAGES
ORAL ANTIBIOTICS (generally 10 days of penicillin V or amoxicillin)	Minimal discomfort	Adherence is usually poor[399] Taste of many formulations unpleasant Paediatric formulations require refrigeration
INJECTED BPG	Guaranteed adherence	Invasive and painful

13. PROVISION OF PRIMARY PROPHYLAXIS

THINGS TO CONSIDER

- What are the barriers to people accessing medical care for sore throats in your setting?
- Are sore throat guidelines being used by all the relevant clinicians?
- How will you know whether the guidelines are being used?

Once guidelines for the diagnosis and management of sore throats have been developed, your programme can focus on delivering primary prophylaxis services. In planning a delivery strategy it can be helpful to think about the demand side (requirements for people accessing services) and the supply side (requirements for of the healthcare staff and system) and barriers which may be confronted by each.[401]

The best way to understand supply and demand barriers in your setting is to ask people about their beliefs and experiences with existing services. The RHD Action Needs Assessment Tool has survey templates to ask people presenting with sore throat about their experiences and to ask health care providers about their knowledge of the disease. These templates can be downloaded to help identify major barriers to primary prophylaxis in your setting.

The following chapter outlines some common barriers to provision of primary prophylaxis and opportunities to address these issues.

> "Two aspects are fundamental to any control programme. Firstly, the people in a community at risk must be aware of the problem, particularly those at high risk who require prophylaxis against recurrences. Secondly, the facilities for preventing the disease at primary and secondary levels must be easily accessible to those who need them."
>
> Edgiton et al, Rheumatic heart disease in Soweto, 1982[402]

Demand-side issues for providing primary prophylaxis

Awareness

Sore throat is a common condition which usually resolves without treatment and rarely causes complications. Many people with a sore throat wait for symptoms to improve and do not seek medical care. For example, in a survey of school children and their parents in Zambia, 30% of participants did not seek treatment for a sore throat. Many who did attempt treatment used only simple home remedies and did not receive clinical evaluation.[403] Similarly, in Tanzania parents reported that they would not normally take a child with a sore throat, with or without fever, to a professional healthcare provider for diagnosis and treatment.[404] In Nepal, of 2245 people attending health facilities 75% knew what sore throat was but fewer than 2% were aware of a link between sore throat and RHD.[405]

Limited awareness of the risks of sore throat and constrained health seeking behaviour were reflected in interviews in at-risk communities in New Zealand in 2013: 'Sore throats are not a priority compared to other needs of children and family/whānau. Their usual response to a child's sore throat is to 'keep an eye' on the child and have them attend school. Medical care was only sought when the condition got worse or coincided with another, more urgent, concern'.[406] More recent work in New Zealand to explore sore throat awareness in at-risk communities indicates that awareness of sore throat and RF is growing – the vast majority of recent survey participants indicated that children with a sore throat need to be seen by a doctor or nurse straight away.[407] High community awareness in New Zealand reflects national prioritisation of RF prevention, discussed further in Box 16, Chapter 14.

Providing education and information about the importance of sore throat treatment is vital for effective delivery of primary prophylaxis. This is covered in detail in Chapter 11.

Accessibility of services

Geographic constraints and logistical issues accessing primary care are common in settings with a high burden of RHD. Cost, difficulty and time of transport to a health facility have been described as barriers to accessing primary prevention in Tanzania.[404] In other settings, access to health care at convenient times (e.g. around the working hours of caregivers) is a barrier to care. This is illustrated by a New Zealand study of an emergency department showing 80% of presentations for sore throat occurred outside the usual opening hours of primary care clinics (after 5pm or during weekends).[408]

Adaptations to the health system to support access to primary prevention have been made in some places. For example, in New Zealand, a system to allow doctors to provide antibiotics for primary prophylaxis directly to patients (without having to visit a chemist or pharmacist) has been developed.[409] In some parts of New Zealand, 6-18 year olds from high risk communities are offered free, 'walk-in' consultations without an appointment with registered nurses for sore throat evaluation and treatment.[410]

In many parts of the world private pharmacies are the main source of medication and health advice.[411] For example, a survey of school children in Nairobi showed that about half of those who remembered having a recent sore throat were treated with medication purchased from local private chemists. Less than 20% had received medication from a dispensary, health centre or hospital.[338] In Zambia, 11% of children with a sore throat had presented to a chemist/pharmacist for treatment.[403] Supporting pharmacies to provide appropriate advice or medication may also improve delivery of primary prophylaxis. In the United Kingdom a pilot study explored whether commercial pharmacies can safely assess sore throat, offer RADT and dispense antibiotics if indicated.[412]

Adherence to antibiotic medication

In addition to accessing health care and being recommended appropriate antibiotic therapy, young people with strep A throat infection need to actually receive medication. Adherence is assured when the administered antibiotic is a single injection of benzathine penicillin G (BPG). However, some guidelines recommend primary prophylaxis with oral tablet antibiotics. The best evidence is for a 10-day course of oral penicillin V twice daily. This difficult to remember, particular when symptoms have resolved.

Adherence to prescribed oral antibiotics can be a barrier to effective primary prophylaxis. In a New Zealand study of 65 people at risk of RF prescribed 10 days of oral antibiotic therapy, only 73.8% of people finished the full course of medication. People who were non-adherent generally said they stopped taking tablets when their symptoms improved.[408] In France, 62% of children with confirmed Strep A sore throat prescribed 10 days of penicillin V three times a day completed the full course of medication.[413] Novel reminders, including sticker charts to help children remember and be rewarded for taking antibiotic tablets have been used in New Zealand to support adherence.[362]

Supply side barriers

Provision of primary prophylaxis requires a functioning health system that is able to procure a stable supply of antibiotics and support individuals presenting for care. Infrastructure, staffing and resources are important determinants of the health system's ability to respond to sore throats. Management of sore throat must also be identified as a clear priority for clinicians to be able to respond to the issue. In one Tanzanian study only 38% of clinicians felt their clinic prioritised sore throat assessment and treatment.[404]

Health care provider attitudes and behaviours

Health care providers must first know that Strep A treatment guidelines exist and be willing to use them. This knowledge and willingness may be influenced by training, education materials and professional experience with RF/RHD.

Even though using clinical guidelines improves quality of care, clinicians may be unaware, unable, unwilling or unsupported to apply them in daily practice. For example in interviews with South African physicians, only 9 of 16 doctors knew that primary prevention guidelines existed, despite the guidelines having been developed over a decade earlier.[414] One study in New Zealand found that only 80% of children with laboratory-confirmed Strep A pharyngitis received appropriate management according to national guidelines.[415] A subsequent New Zealand study identified improvements in prescribing antibiotics according to national guidelines but ongoing variation in the recommended duration of therapy.[408]

Strategies for improving use of guidelines may include:

- Develop local guidelines in consultation with local clinicians. Engaging clinical leaders and professional societies makes it more likely that clinical colleagues will change their practice.
- Utilise formats that are accessible to target audiences – web-based if the Internet is available, hard copy for distribution to remote locations, or mobile phone applications.
- Publish a summary of the guidelines in a journal, newsletter and health related magazines, hospital and general practice newsletters and other media.
- Disseminate guidelines at conferences, medical and nursing schools and at meetings and seminars.
- Ask clinical groups, specialist colleges, public health authorities and professional bodies to endorse the use of the guidelines.
- Integrate guideline recommendations into continuous quality improvement processes. Support clinicians to audit clinical practice against guideline recommendations.

An additional barrier to the use of sore throat guidelines may also be clinician concerns about adverse reactions to intramuscular BPG injections. Details about BPG and support for providers to deliver safe injections is outlined in Chapter 17.

Medication procurement

Ensuring adequate supplies of medication are available for primary prevention is another important role for the health service. Procurement of BPG and other essential medicines is outlined in Chapter 17.

14. ACTIVE CASE FINDING (SORE THROAT SCREENING CLINICS)

THINGS TO CONSIDER

- Are other school based health services delivered in your area?
- Could sore throat management be included in the role of school nurses or health workers?
- What capacity is there to provide reliable screening services and act on outcomes?
- Are there other novel ways to deliver sore throat management in your communities?

Even when all the components outlined in Chapter 13 work perfectly some sore throats will be too mild for families to seek treatment or barriers to access will be prohibitive. Screening or treating sore throat and Strep A infections in school children is an attempt to address these issues and maximise the impact of primary prevention.[416] A small number of historic sore throat clinics were run in the United States.[417,418] Intermittent opportunistic school-based screening for Strep A throat infections occurs as part of an RF/RHD outreach programme in Kenya – reaching 27,661 students between 2008-2012.[187] The most significant contemporary experience with sore throat clinics has occurred in New Zealand.

The school-based sore throat management programme in New Zealand is one of the largest interventions to address rheumatic fever ever conducted - involving nearly 55,000 children and $23 million NZ dollars. Capacity to extrapolate this programme to other settings, particularly in low and middle-income countries, is unclear. However, the programme is of considerable international interest and is presented here as a foundation for discussion about the role of school-based sore throat programmes.[419]

Dr Duncan Matheka examines a young patient for a sore throat.

BOX 16:
The New Zealand Experience

A randomised control trial involving 22,000 school children was carried out in South Auckland, New Zealand between 1998 and 2001.[6] South Auckland is an endemic region of disease with a large Māori and Pacific Island population at greatest risk of rheumatic fever (RF incidence 60/100,000/year). Schools were randomised to receive a school based sore throat clinical or standard treatment. The intervention group was diagnosed and treated for GAS pharyngitis by nurses in a school-based clinic programme. Treatment with a 10-day course of twice-daily oral penicillin, administered under nurse supervision, was initiated after a positive throat culture. Community health workers made daily classroom rounds to ask consented children if they were experiencing symptoms of sore throat and performed monthly throat examinations to actively find asymptomatic cases.

The endpoint of this RCT was the incidence of RF determined by the Jones Criteria with the additional use of echocardiography to screen for carditis. The control group received standard general practice care outside of the school setting. Results showed a 20-30% relative risk reduction in RF cases in the intervention group compared to the control group. However, these findings did not reach statistical significance.

On the basis of these results no recommendation could be made to advocate for the implementation of school-based Strep A pharyngitis diagnosis and treatment programs as a way to decrease incidence of RF. It has since been suggested that the New Zealand RCT did not reach significance because of household contacts.[419] Specifically, management of the intervention group did not include swabbing of symptomatic sibling who may have been in the control group.[412]

A subsequent meta-analysis of school-based studies from New Zealand, Cuba, Hawaii, inner-city Baltimore and two Indigenous communities in America did, however, find a significant 60% reduction in incidence of ARF conferred by school-based sore throat clinic programs compared to general practice care.[422] The New Zealand study was the only RCT included in this meta-analysis, other published data was low quality observational data.[423,424]

In 2010 and 2011 RF became a political issue in New Zealand, driven by rising rates of RF and the disparity in incidence for Māori and Pacific Island communities compared in non-Indigenous New Zealanders.[11] A Rheumatic Fever Prevention Program (RFPP) was established in 2011 to prevent and treat strep throat infections, which can lead to rheumatic fever. The programme was expanded significantly from 2012 following the introduction of the five-year rheumatic fever Better Public Services target. Specifically, the target was to 'reduce the incidence of ARF by two-thirds from a baseline of 4.0 per 100,000 in 2009/10–2011/12 to 1.4 per 100,000 by 2017.'[40] The government invested about $65 million to identify and trial new initiatives to reduce the rheumatic fever rates throughout New Zealand. The RFPP was delivered in 11 District Health Board regions with a high incidence of rheumatic fever.

The RFPP had 3 main strategies:[40]

- increase awareness of rheumatic fever, what causes it and how to prevent it.
- reduce household crowding and therefore reduce household transmission of strep throat bacteria within households.
- improve access to timely and effective treatment for strep throat infections in priority communities. This included both school-based and primary care sore throat management and primary care sore throat management.

The school-based component of the RFPP was informed by recommendations of the New Zealand Heart Foundation and other professional bodies based on the 2009 meta-analysis of school sore throat screening.[40]

A school-based sore throat management programme was implemented in 10 of the 11 DHBs with a high incidence of rheumatic fever. It is important to note that a number of DHBs had a school-based sore throat management programme before the implementation of the RFPP, other scaled up their programmes rapidly.[406] By early 2014 the programme peaked in intensity including 251 schools, covering 53,998 children in high risk communities for school based throat swabs.[40] Each region delivered the school programme differently, depending on resources and what worked within their populations. However, for all schools, children found to be positive for Strep A on throat swab received free antibiotics without having to fill a prescription. Some schools also included phone calls to support antibiotic adherence and management of skin infections. In combination about 36% of high risk children had access to school sore throat programs in 2014 covering 40 weeks of the year during term time.[40] School holidays mean that school-based programmes do not reach high-risk children throughout the whole year.

A number of evaluations and analysis of school-based elements of the RFPP have been conducted. An Implementation and Formative Evaluation was commissioned covering the period July 2011 - December 2012.[406] This evaluation provided detailed information about the initiation of the RFPP including input from a range of key informants. Themes included the need for consultation, integration and sustainability planning. Practical issues about service delivery models were addressed. The findings from this evaluation were incorporated into further development of the RFPP. Scientific feedback on the efficacy of a school-based throat screening was addressed. Recommendations for strengthening the programme in 10 domains were identified.[406]

In 2015 an Interim Evaluation was commissioned to outline the efficacy of the school-based sore throat programme. This report concluded that RF rates were decreasing in New Zealand but that school-based services alone would not achieve the Government target to reduce RF by two-thirds by 2017. [40] Specifically, the analysis of the effectiveness of the school-based services in the 10 DHBs that have implemented this service showed an overall decline of 17% in acute rheumatic fever cases attending schools offering the school-based sore throat management service. This decline was not statistically significant. For one large DHB, the decline was larger at 31% but was also not statistically significant. Cost effectiveness analysis within this evaluation informed the decision of some DHBs to discontinue school-based programs and manage sore throats through primary care.[419] An academic analysis of the RFPP school-based sore throat programme will be published in 2018.

New analysis suggests measuring changes of RF incidence in New Zealand is an ongoing challenge.[426]

RF was retired as a Better Public Service target in June 2017. However, rheumatic fever prevention will continue to be a focus for the 11 DHBs with a high incidence of rheumatic fever. The government has allocated funding till 2022 for the 11 DHBs so they can continue to deliver a balanced mix of rheumatic fever prevention activities to address rheumatic fever and reduce rheumatic fever rates.

The totality of the programme (including sore throat management, primordial prevention and largescale community engagement and participation) have been described as 'an exemplar in relation to integrating primordial and primary prevention activities across government departments to communities'.[427]

BOX 17:

The challenge of low sore throat reports

The relationship between RF and preceding sore throats is complicated - a significant proportion of children present with RF without any recollection of sore throat. This is a challenge for primary prevention programs, particularly large scale throat swabbing programs. For example:

- In the United States of America an eight year study was conducted to explore increased incidence of RF. Over that period only 28% of children with confirmed RF reported a history of a sore throat and only had 17% sought medical treatment for that pain.[129]
- In Pakistan in the 1980s only 30 - 40% of children had a strong history of sore throat preceding RF.[428]
- In a contemporary New Zealand setting only 46% of children reported a sore throat prior to RF.[429] In another New Zealand study 14 of 19 children recalled a sore throat within 63 days of an episode of RF.[420]
- In Australia, 33% of all children with ARF reported a recent sore throat, reducing to 25% for Aboriginal and Torres Strait Islander children living in remote communities.[430]

Poor correlation between sore throat and RF may represent recall bias, asymptomatic infection or Strep A infection from a skin source (see Box 6). Clearly, the potential effectiveness of primary prevention is limited by the proportion of ARF cases that are preceded by a sore throat sufficiently severe to lead to a presentation to a health facility. If symptomatic Strep A sore throats are not the primary driver of cases of RF, active case finding for pharyngitis mechanisms may have limited impact on the burden of disease. This may be further confounded by pharyngeal carriage (see Box 14).[40]

Challenges in adapting school based-sore throat programs

The cost and logistical challenges of introducing school-based Strep A screening programmes in low resource settings are likely to be prohibitive without considerable modification of the New Zealand approach.[424] In Australia, a high resource neighbor to New Zealand, there may be scope to explore school based primary prevention. However, a review of Australia's rheumatic fever strategy noted "The geographical isolation of many of the Indigenous communities will make the implementation of a similar intensive primary prevention intervention far more challenging and costly than that in New Zealand. The conclusion of the evaluation team is that implementing a national strategy such as New Zealand's across Australia would not be appropriate at this point in time. However, piloting the approach in a small number of communities is worth considering."[431] The New Zealand experience demonstrates that engaging communities to identify locally workable approaches to reduce is critical to success.

- School attendance in countries with a high burden of RHD is often low and is likely to be lowest in some of the children at greatest risk of RF, particularly girls from low income families, Indigenous communities or refugee/ migrant communities.
- An inconsistent number of children report sore throats preceding RF (see Box 17) representing a variable opportunity to for primary prevention.
- Understanding of Strep A carriage is incomplete; children with Strep A positive throat swabs will not necessarily have pharyngitis from Strep A (See Box 14).
- Costs of delivering care may be very high, particularly if population is dispersed in rural and remote locations.[197]

15. STREP A VACCINE DEVELOPMENT

THINGS TO CONSIDER

- How can your RHD Control Programme amplify global calls for a Strep A vaccine?
- Do you have enough information to advocate effectively for development or adoption of a Strep A vaccine?
- What other information may be needed to plan for vaccine delivery and implementation?

A vaccine against Strep A offers promise for definitive control of Strep A infection and their consequences, including RF and RHD.

Attempts to develop a Strep A vaccine have been underway since the early 1920s and a number have progressed to early human trials.[432] Progress towards a safe, effective, affordable and practical Strep A vaccine has accelerated in recent years, including signals of prioritisation from the World Health Organization and the International Vaccine Institute.[433] A clinical trial pathway has been developed to support a pathway to licensure for candidate vaccines.[434] Development of a commercially available vaccine is still some years away – necessitating an ongoing focus on comprehensive RHD control programmes – but is expected to be achievable with sufficient resourcing and political will. However, scientific development of a Strep A vaccine is only part the process required for a future vaccine to be used prevent RHD. In addition to formulation and licensure, countries must be willing to use the vaccine, assume any associated costs and have a robust system to deliver the vaccine to people in greatest need.

Comprehensive RHD control programmes can have an important role supporting Strep A vaccine preparedness. Countries, communities and control programmes are the primary stakeholders in vaccine development. Local engagement is critical for producing a vaccine which is needed, accepted and adopted. RHD control programmes have the best possible insight into why a vaccine is needed and are critical stakeholders in vaccine planning and advocacy. This chapter outlines the opportunities RHD control programmes have to contribute to Strep A vaccine development and delivery.

Vaccine awareness and advocacy

Even when vaccines exist they may not be used unless a country has a high burden of the disease and prioritises control. For example, a vaccine against Haemophilus influenzae type b (Hib) was available in developed countries in the year 1990. However, by 2015 only 13 of 75 developing countries eligible for free Hib vaccine supplies from the Global Alliance for Vaccines and Immunisation (GAVI) had included the new vaccine on their national immunisation programmes.[435] Complex barriers to vaccine uptake include perceptions of need, political prioritisation, beliefs about safety and efficacy, strength of scientific recommendations and health system capacity.[436,437]

A number of resources have been developed to help countries plan for vaccine implementation, even before a licensed vaccine becomes available.[436,438] One of these resources, developed by the World Health Organization, focuses on three domains: the disease, the vaccine and the strength of the immunisation programme and health system.[438] This resource poses five main questions about the disease which may influence the decision to adopt a new vaccine. These questions are outlined in Table 16 alongside the potential role of RHD control programmes in providing answers.

Table 16: Key questions on disease characteristics for countries considering vaccine introduction adapted from 'Principles and consideration for adding a vaccine to a national immunisation programme – from decision to implementation and monitoring'.[438]

Does the disease cause significant disease burden?	RHD is one of the largest and most significant outcomes from Strep A infections. Therefore, documenting the burden of RHD is critical for informing decisions about introducing a Strep A vaccine in low resource settings. There is good evidence that locally measured data on burden of disease has a greater impact on decision makers than international estimates.[439,440] Approaches to collecting burden of disease data are provided in Chapter 1 and are an important element of RHD control programmes.
Does preventing the disease contribute significantly to the goals and align with the priorities established in the national health and development plans?	Pre-existing national priorities inform vaccine adoption decisions. Therefore, advocacy efforts to ensure RHD is included in service plans, non-communicable disease plans and other strategic documents is particularly valuable. This advocacy can facilitate service delivery and resourcing in the near term (for comprehensive RHD control programmes) and in the future as a Strep A vaccine becomes available. Approaches to advocacy and government engagement are outlined in Chapter 6.
Is the disease perceived to be important to the public and the medical community?	The more visible and important the disease is to the community, the greater the acceptance of and demand for the vaccine will be.[438] Perceptions of disease burden, severity and treatment are an important determinant of vaccine adoption decisions. In the context of RHD this means that people living with RHD and the wider community need to be aware of the disease and it's impact. Communities can partner with clinicians to call for action to address the disease. Community education activities are outlined in Chapter 11. Ultimately, vaccine adoption and other disease control decisions reflect political priorities – which can be informed by the experiences of people affected by the disease.
Is the vaccine recommended by WHO and is control of this disease in line with global or regional priorities?	Global advocacy is underway at the highest level of WHO to ensure that RHD is identified as a priority condition for prevention.[433]
Does preventing the disease contribute to improving equity among socioeconomic classes and population groups?	RHD is most common in the most vulnerable groups, including people living in greatest poverty, Indigenous people, refugees and those with limited access to health services. Defining and describing inequalities in the distribution of RHD may provide a useful rationale for introduction of a Strep A vaccine. Outlining the burden of disease in sub-populations is addressed in Chapter 1.

"Although the burden of disease was clearly necessary for adoption decisions, it was not generally sufficient; political prioritisation was also very influential."

Burchett et al, New vaccine adoption: qualitative study of decision-making process in seven low and middle income countries, 2012[439]

SECONDARY PREVENTION

Secondary prevention is based on case finding, referral, registration, surveillance, follow up and regular secondary prophylaxis for RF and RHD patients.[441]

"Secondary prophylaxis is the continuous administration of specific antibiotics to patients with a previous attack of rheumatic fever, or well-documented rheumatic heart disease. The purpose is to prevent colonisation or infection of the upper respiratory tract with group A beta-hemolytic streptococci and the development of recurrent attacks of rheumatic fever."

A WHO Expert Consultation on RF and RHD, 2001[441]

Secondary prevention is an effective way of slowing or preventing the progress of rheumatic heart disease from mild valve changes to advanced valvular heart disease. A low level of penicillin in the blood prevents Strep A infections, which in turn prevents the recurrent episodes of RF. Prevention of RF recurrences is strongly associated with better cardiac outcomes.[442,443] Some emerging data suggests that a high level of regular penicillin administration may reduce deaths from RHD.[444]

Delivery of secondary prophylaxis generally requires an 'RHD register' which records the names and details of people who should be receiving regular antibiotics. These are often called 'register-based RHD control programmes'. The World Health Organization (WHO) and the World Heart Federation (WHF) endorse register-based RHD control programmes to reduce the burden of RF and RHD.

16. THE RF/RHD REGISTER

THINGS TO CONSIDER

- Do you have an RF/RHD register?
- Where do you store the RF/RHD Register, and how do you maintain it?
- How are people with RF/RHD added to, or removed, from the register?
- How do you ensure patients' confidentiality in your register?
- How is your register data shared with different collaborators and groups?
- What is the aim of establishing your RF/RHD Register?

What is a register?

A disease register is a list of people who have been diagnosed with, or are suspected of having a disease.[441] RF/RHD registers are used for collecting data about people living with RF/RHD and some of their clinical details.

The concept of RF/RHD registers was first introduced in the United States in the 1950s. These registers helped provide newly-developed regimens for secondary prophylaxis, and contributed to the declining burden of RHD in the USA.[445]

By the 1970s, WHO endorsed the register-based approach, rendering it to be an essential part of RHD control. Further, the Pan-African Society of Cardiology (PASCAR)-led 'Awareness Raising, Surveillance, Advocacy and Prevention' (A.S.A.P) approach has set registers to be one of the four essential pillars for RHD control.[446] Register-based programmes have assisted with the provision of prophylaxis in many communities across the globe.

Why is a register so important?

An RF/RHD register can assist with routine assessment and surveillance, recording of prophylaxis delivery, the recall of patients who are due for or miss doses of BPG, and finally, improve health education and health promotion programmes. On a global scale, registers provide information about the burden of disease. However, data quality depends on the quality of register management. An overview of the benefits of registers, for both people living with RF/RHD, and for health systems, can be found in Table 17.

A contemporary study from New Zealand demonstrated the incremental value of register-based care. In this study, young people receiving register-based secondary prophylaxis injections were able to receive 94% of their injection. In contrast, those who received their injections through an unstructured primary care programme received only 37% of their scheduled injections.[447]

To attain maximum benefit from register-based programmes, the register must be user-friendly. This means that staff who update the register and use the information should be involved in register design and scaling up.

A review of different kinds of disease registers in African countries found that 'Externally-led efforts can take away a sense of ownership within the health system, result in duplicate data collection, and often increase system fatigue. Frontline health workers may not use registers they found unsatisfactory, and local health authorities may not endorse registers that fail to meet their needs. These case studies demonstrate that register systems inspired by grassroots solutions are often more accepted and more likely to be successfully scaled.'[448]

Table 17: Benefits of RF / RHD registers

FOR PEOPLE LIVING WITH RF/RHD	FOR HEALTH SYSTEMS
• Improves delivery of consistent, disease altering, secondary prophylaxis through recalls and reminders to have secondary prophylaxis. • Facilitates priority-based care and clinical review.	• Helps to identify people with poor adherence for additional support. • Provides information about the burden of disease over time. • Facilitates monitoring of recurrence rate and other indicators. • Provides a monitoring tool for data quality in research.

Establishing a register

Minimum data set

Global consensus is still emerging about what kind of information should be collected and stored on an RHD register. The WHF has previously developed a register data collection form and template.[449] Some national programmes have established their own data collection instruments and standards.[450] PASCAR is also working to establish a minimum data set for RHD registers, according to the aim of work, e.g. primary tool for clinical management, or research activities.[451]

Registers targeting combined endpoints for clinical and research outcomes are achievable, however, the amount of data required can be overwhelming for frontline health staff to collect and manage. [448] Therefore, you should carefully consider the goals of your register and subsequent data use during the design phase of the project. Consultation with international colleagues and programmes may be required to help with decision making regarding data collection choices.

Ideally, registers should be as inclusive as possible, including paediatric and adult patients and the full spectrum of disease severity; i.e. registers should include data about history of RF, screening-detected asymptomatic RHD, people actively receiving secondary prophylaxis, people who are awaiting or have undergone surgery.[328] Such a comprehensive approach offers scope for improved clinical care and valuable epidemiologic insights about the natural history of disease progression.

Privacy, confidentiality and data security

Use, ownership, patient confidentiality and handling health data are complicated issues worldwide. [445] Many settings have struggled to establish protocols to manage confidential health information. Privacy requirements for RHD registers may impact the decision on which data is collected, who can view it and how the data can be used.

Compliance to local laws, standards and procedures must be considered while establishing a register. It is crucial to seek advice and consult the relevant bodies before establishing an RHD register – you may need to seek input from ethics committees, health authorities, and/or other register-based programmes.

Training

Staff will require training to use and maintain the RHD register. The more complicated the system is, the more time your team will take to become familiar with it. Using a new system to build the RHD register may require special training, thus, integrating the RHD register with the existing systems at your facility will help your staff to be more familiar with the register.[441]

In addition to frontline health workers, maintaining the register usually requires dedicated staff time to review information, follow up incomplete data, respond to requests for information and provide reports to key stakeholders.[448]

Register logistics

Electronic registers have shown efficacy and accountability in terms of data entry, revisions, and management. These benefits are possible from any computer or smart device at any site depending on the compatibility of the system you use for your RHD register. Ideally, your electronic register will integrate with local electronic health information systems. However, countries with a large burden of RHD tend to have a poor health information infrastructure, and it is often not possible to begin with a 'perfect' solution. Starting with a smaller register of local patients may provide a foundation for expansion.

Early stage registers require two initial practical decisions: location and format. Information to inform these decisions is outlined in the following paragraphs, with advantages and disadvantages of each approach summarised in Table 18.

Table 18: Considerations for register location

	ADVANTAGES	DISADVANTAGES
LOCAL	• Useful for following up individual people living with RHD. • Can be updated by local clinical staff. • Helps in filtering the recorded data, and accordingly, send precise data to its target pool.	• Can be difficult to transmit information to a central register therefore has limited value as an epidemiologic tool. • Usually requires using the resources on site. • Requires proper planning for data sharing.
CENTRAL	• Provides an overview of the burden of disease among the population. • Valuable for mobile populations or people who move frequently.	• Requires systems to provide information back to the health system level. • Usually include basic data, which may not be clinically relevant for primary care provision, or research. • Requires strong data management and revision protocols to ensure data accuracy as received from the local registers.

Table 19: Considerations for register format

	ADVANTAGES	DISADVANTAGES
PAPER	• Less training is required. • Data collection is easier especially in busy clinics.	• Data is difficult to extract and analyse. • Data may be lost or misplaced. • Requires a dedicated physical storage space. • Hand written data can be misunderstood. • Calculations of scheduled visit dates is subject to human error.
ELECTRONIC	• Reports and data easily extracted. • Supports integrative care if combined with electronic health records. • Automated calculation and setting of follow up schedule programmes.	• May requires special technical setting. • May be more expensive to establish and maintain. • Staff training is likely to be required. • Subject to computer-related problems, e.g. system failure or bugs.

Register location

Central registers

Centralised registers are maintained in major hospitals or online. Centralisation can support the provision of prophylaxis for people who move between different regions or health clinics. For example, 30% of people receiving secondary prophylaxis moved within New Zealand or overseas over an eight-year period.[447] The collection of information from more areas can also provide valuable epidemiological data to assist public health clinicians, managers and policy makers to understand the disease burden in different areas and assist with allocating the resources to where they are needed most.

In some places, local register data may be shared partially with the central register. For example, in Nepal local registers record patients admitted with RF/RHD, and share summary data with the national (central) register maintained by the Nepal Heart Foundation.[327]

Local registers

A local register can be a paper or electronic list of people living with RHD in the community. There is some evidence that diagnosis and management of RHD occurs in clusters around local register sites; so decentralising screening and data collection to local registers may improve coverage to a wider populalation.[452]

Register format

Paper-based registers

Patient data is recorded in a book or on paper cards and stored in cabinets where they are arranged in a way that suits the local situation. For example, cards can be arranged according to the month in which the person requires their next appointment.

One of the challenges of paper registers is ensuring that data is stored safely and protected against loss or damage. For example, in Samoa, a misplaced paper register reduced the effectiveness of the RHD control programme: 'This manual register was well kept from 1984 to 2002 but unfortunately this register was lost. The rheumatic fever work and clinics continued with less coordination between the various centres in both islands looking after rheumatic fever patients.'[453]

Paper-based registers have some other down sides. Data is hand-written and, if not written clearly, can be misunderstood. Also, the manual calculation of follow up visits or next scheduled injection may be prone to errors, especially when compared to automated, computer-based calculations.[45] See Table 19 for an overview of the advantages and disadvantages of paper-based registers.

Computer-based registers

A computer-based register may be a unique database used only for RF/RHD management and control or one that is part of a broader patient information system that is used for all patient management. It is important that a computer-based register has the fields to store the required information, that staff can easily use it and that it can generate a list (a recall list) for staff to use as a guide as to who is due or overdue for their prophylaxis or clinical review.

Although computer registers are more durable than paper copies (and more easily backed up) software and hardware maintenance is required. This should be factored into your budget. See Table 19 for an overview of the advantages and disadvantages of computer-based registers.

Different kinds of electronic registers are available.

- The simplest form is an electronic spreadsheet, using dedicated software processors. The most famous of them is commercial Microsoft Excel, and open source Apache OpenOffice Calc. Many people are familiar with spreadsheets as they are relatively easy to use. However, version control between different copies of the document may be difficult and may prohibit data input from multiple users.
- Standard database software can be used to develop a register. For example, the WHF developed a Microsoft Access database for collecting RHD information in the early 2000s.
- A number of research projects have used REDCap databases to collect data, including studies in Uganda, Nepal and Brazil.[49,454,455] REDCap is a free, secure, web-based platform that offers easy design of data collection instruments, data capture and management, and data export to multiple electronic formats including spreadsheets. The REDCap consortium offers strong support to their users and regular updates for the system.
- Another example is 'eRegister for RHD', which has been developed using CommCare register software; this is used by the RHD programme in Zambia.[456] CommCare uses data collection forms that can later be extracted as a spreadsheet. Data input occurs through laptops, tablets and smartphones and is stored in an electronic cloud. PASCAR is in the process of expanding this technology for use throughout the region including development of an eRegister app.[457]
- Some programmes choose to use custom software for their RHD register. For example, Fiji has invested in a Rheumatic Fever Information System (RFIS) which was developed by a local software company.[458]

Maintaining information flow to and from the register

"The effectiveness of a register based program depends on the accuracy of the database, how well it is maintained and how well the information is disseminated."

Eissa, et al, Assessment of a register-based rheumatic heart disease secondary prevention programme in an Australian Aboriginal community, 2005.[459]

The RF/RHD register is only as good as the information entered. Complete, high quality data is critical for programmatic success. You will need a clear plan for ensuring people can be added to the register. There will likely need to be multiple entry and status change points within the register. These are outlined in Table 20.

BOX 18:
Expect an apparent increase in cases when notifications and registrations begin

Starting or strengthening a control programme tends to increase health worker and community awareness about RF. This may make people more vigilant, prompting notification of suspected cases of RF, which may have been overlooked.[47,460] A surge of interest can sometimes make it appear as though there is a new epidemic of disease.

"The first months of the programme led to a 10–20% increase in the number of rheumatic fever cases admitted to hospital, because of the renewed attention paid to the disease."

Bach et al, 10-year educational programme aimed at rheumatic fever in two French Carribean Islands, 1996.[34]

Sr Mere Dula, Registered Nurse from the Fiji Rheumatic Heart Disease Control Program with the ARF and RHD Patient Register Book.

Table 20: States for RHD Registers

POTENTIAL ENTRY POINTS TO THE RHD REGISTER	POTENTIAL CHANGES TO REGISTER STATUS
1. AFTER AN EPISODE OF RF People should be entered into the register at the earliest possible opportunity – the first episode of RF. This requires clinical staff in primary and secondary care to know how to diagnose suspected/confirmed RF, contact details and other information required for the register. In addition to the date of clinical presentation, the date of first symptom/complaint should be recorded. It is important to encourage clinicians to contact the register about all cases of RF – suspected and confirmed, first episode and recurrences. Such processes reduce the decision making burden for individual clinicians and provide as much information as possible to your programme. Information about recurrence can also be added to individual clinical records. Clinical review of cases reported to the register may provide valuable support to primary care clinicians and improve data quality.	**1. PROPHYLAXIS PLAN CESSATION OR COMPLETION** Treatment and management guidelines should be clear about the duration of secondary prophylaxis – this may be guided by the priority management categories (Chapter 19). The patient's history of RF and the presence of RHD-associated heart valve affection will guide the decision to cease secondary prophylaxis. Prophylaxis strategies, including BPG injections or other, should be ceased when it is clinically appropriate. Monitoring systems are required to ensure revision of a prophylaxis plan once treatment is complete. The decision by specialist clinicians to stop treatment needs to be clearly communicated to teams responsible for administering secondary prophylaxis. In a review of the Australian Northern Territory programme, two patients continued to receive secondary prophylaxis, despite clinicians deciding BPG was no longer required.[459] The register may also be able to document whether the planned duration of secondary prophylaxis was delivered accurately, according to local guidelines; whether secondary prophylaxis was stopped early following expert clinical review, or the prophylaxis was stopped without clinical consultation.
2. AFTER FIRST PRESENTATION OF SYMPTOMATIC RHD Clinicians who diagnose RHD need to be able to contact the register coordinator to enter people living with RHD into the register. Advanced cases of RHD may be identified late in adults, and rarely in the elderly. Therefore, adult clinicians, midwives, and primary care staff will need to know to contact your RHD control programme. Ideally, clinicians should be able to contact the register to check, update, or confirm clinical information – including delivery of secondary prophylaxis, planned follow up, referrals for specialist review or surgical evaluation.	**2. INACTIVE/DROP OUT PATIENTS** All programmes are subject to lose some patients to follow up – due to unreported deaths, unplanned travel, unplanned changes in contact details, or active avoidance. These patients continue to be relevant epidemiologically, even if secondary prophylaxis cannot be delivered. The removal of data completely from a register will limit the ability of the control programme to report epidemiological findings. An 'inactive' category allows retaining data without active care delivery; therefore, you will need to define 'lost to follow up' category in your register.

POTENTIAL ENTRY POINTS TO THE RHD REGISTER	POTENTIAL CHANGES TO REGISTER STATUS
3. AFTER ECHOCARDIOGRAPHY SCREENING ACTIVITIES Echocardiographic screening research (outlined in Chapter 20) should be linked to the RF/RHD register, in order to ensure that the screening-identified RHD patients are receiving appropriate care and follow up. (Conversely, if your register has been initiated as part of an echocardiography screening process you should consider making it possible for other people in the same community diagnosed with RHD to be added to the register).	**3. DEATH** Recording deaths of individuals on the RHD register is important in order to: • Avoid inappropriate distress for families and communities by attempting to follow up deceased individuals. • Ensure that resources are not consumed attempting to follow up deceased patients. • Understand the mortality burden of RHD, and explore the burden particularly if details about cause of death are available. • Add to records of surgical outcomes where appropriate (see Chapter 24). In some places, it may be possible to access hospital death records to identify RHD patients who have died.[317] Primary care clinicians, midwives, hospitals and communities can be encouraged to contact the programme about people who have died while on the RHD register, or receiving secondary prophylaxis. Detailed recording of data related to the cause(s) of death is crucial in maintaining an RHD register. (See also Chapter 21 – mortality and palliative care).
4. TRANSFER IN FROM ANOTHER PROGRAMME In some countries, RHD patients are from mobile communities who move frequently for work, healthcare, family or traditional responsibilities.[447,459] You will need to set an accurate protocol for accepting registrations from other programmes and for entering people who arrive unexpectedly seeking secondary prophylaxis. These may include refugees and new immigrants moving from areas with a high burden of RHD and limited health infrastructure.[430] Other sources of information for the register may include: notifications, hospital discharge records, clinical letters, echocardiogram reports or professional correspondence.[317] These records may also provide valuable information about the clinical status of people already registered in the RHD register.	**4. TRANSFER OUT TO ANOTHER PROGRAMME** Patients entered in your register may need to move outside of your programme. When travel or relocation is planned, you should identify a new provider of secondary prophylaxis and follow up. Options may include: • Giving people on the register a copy of a referral note and medical information prior to relocation. • Providing a card with the name and contact details of your programme to be presented at other hospitals or health providers as needed. • Contacting other RHD control programmes or care providers in the intended destination prior to travel. Consent to share clinical information with other programmes should be obtained.

Closing the register

The goal of comprehensive register-based RHD control programmes is to reduce the burden of RHD. The WHF has an international goal 'to achieve a 25% reduction in premature deaths from RF and RHD among individuals aged less than 25 years by 2025'.[227]

Although the thresholds for disease elimination and control at national and regional levels are still under development, it is reasonable to expect that comprehensive programmes will see RF/RHD recede as a public health priority. An 'exit strategy' for how and when to close registers is important for planning. This can be achieved by identifying criteria for closing registers in advance to avoid premature closure.

The best information about closing/phasing out register-based programmes comes from the United States in the 1970s.[462] In 1977, twenty-nine states had some kind of RHD register, and by the year 1979, only 11 of these states had ongoing register-based programmes. Reduction in programme numbers was attributed to an apparent decrease in RF diagnosis, and budget constraints.[462]

A similar experience was noted in New Zealand:

> "The recent discontinuation of the Waikato register shows that previous longevity does not assure continuity of register-based programmes. The Rotorua experience in the early 1990s suggests that, despite local leadership, these programmes may be vulnerable to the effects of health sector restructuring."
>
> Thornley et al, Rheumatic fever registers in New Zealand, 2001.[463]

Retaining some mechanism for monitoring and supporting care for RF/RHD is advisable, even when disease control targets have been met. A background rate of disease persists in high resource settings and disease resurgence after a period of control is possible. For example, after the burden of disease had fallen, in 1985 Utah experienced an eight-fold increase in RF diagnosis.[129] Although this cluster of patients had some atypical features, the outbreak demonstrates the importance of maintaining some mechanism for identifying and managing RF and RHD.[129] Similarly, in the United Kingdom ongoing sporadic cases of RF were detected in the late 1990s.[464]

17. BPG AND OTHER ESSENTIAL MEDICINES

THINGS TO CONSIDER

- Is BPG on your national essential medicines list or formulary?
- Do you have stock outs or shortages of BPG?
- How is BPG use recorded so that new product can be reordered?
- Do you have a system for ensuring all people are asked about a history of penicillin allergy?
- Do you have guidelines for management of anaphylaxis?
- Do you have access to adrenaline and other treatments for anaphylaxis?
- Do you have mechanisms for monitoring or reporting adverse drug reactions?

Antibiotic medications are needed for primary and secondary prevention of RF. In primary prevention they are used to treat Strep A infections, and in secondary prevention they are used to prevent new Strep A infections causing recurrences of RF. Securing a reliable, high quality supply of antibiotic before beginning a secondary prophylaxis programme is important – otherwise people on the register could be exposed to the risks of intermittent antibiotic therapy (painful injections, allergy and inconvenience) without the continuous supply necessary for significant benefit.

Other medications are needed for relief of symptoms from RF and RHD. Medications may also be used to reduce the risk of complications from RHD, including anticoagulation discussed in Chapter 22.

The RHD Action Needs Assessment Tool offers a useful survey template for independent dispensaries. This questionnaire explores access to a number of essential medicines in a specific location.

Benzathine Penicillin G

The antibiotic benzathine penicillin G (BPG), also known as benzathine benzyl penicillin, is the product most commonly used for primary and secondary prophylaxis. BPG is injected into muscle (intramuscular injection) and provides a peak of penicillin concentration in the blood before dispersing very slowly. This ensures that penicillin levels are detectable in blood for a number of weeks. This low level of penicillin is thought to protect against recurrent Strep A infections, providing protection for secondary prophylaxis.[465]

Globally there are two formulations of BPG.[466] A pre-mixed liquid formulation which is produced under patent by a single manufacturer, dispensed in a prefilled syringe and is relatively expensive. The liquid formulation requires refrigeration and is most widely used in high income settings. Powdered forms of BPG are produced by a number of different generic manufacturers and are inexpensive.[467] The powdered forms must be mixed with a sterile diluent (usually water or normal saline) prior to injection. It does not require a cold chain and can be stored for a number of years. TIPs focuses on powdered formulations of BPG which are used in most settings with an endemic burden of RHD.

Supply and stockouts of BPG

Stock outs and shortages of BPG have been reported worldwide since the early 2000s.[466,467] In 2016 a detailed analysis of BPG stockouts, funded by the Bill and Melinda Gates Foundation, was conducted by the Clinton Health Access Initiative (CHAI) and the WHO. This review focused on the use of BPG for the treatment of syphilis between 2014 and 2016 but findings are applicable to any use of BPG. Forty one percent of the 95 countries surveyed reported a BPG shortage.[467] Contributing supply and demand side issues were identified during this study and are summarised in Table 21.

Table 21: Contributors to BPG stockouts (summarised from[467])

SUPPLY SIDE	DEMAND SIDE
• BPG is a low-cost product which means low profit margins for manufacturers. Some countries have a maximum price set for BPG which exacerbates this problem. Many manufacturers have left the market because BPG is not profitable. • It is difficult for new manufacturers of BPG to enter the market because producing BPG requires specialised equipment which cannot be easily shifted to make other drugs. • Purchase orders from countries to buy BPG are often erratic and small volumes which makes it difficult to plan production schedules. Despite these issues a global review found that BPG manufacturers have capacity to scale up production to meet need. Demand side barriers are the major drivers of BPG shortages.	• Difficulty forecasting demand for BPG. This can occur when information on BPG use is not passed on from clinics or when orders are based on historic patterns of use which may have coincided with a period of stock out. This creates a spiral of misinformation when previous shortages lead to underestimates of future demands. • Inflexible purchasing cycles – in some places drug supplies are only ordered annually. Given the long lead times to receive BPG this can mean long delays between ordering BPG and receiving the product. • Limited funding to purchase BPG. • Limited registrations for BPG products which makes it difficult to switch between different brands of the product during periods of stockout. These issues are amplified by substitution behaviours during stockouts when healthcare workers may change to oral antibiotics temporarily. Although less effective these are often more acceptable and are then perceived as a permanent substitute.

Practical steps to reduce the risk of shortages and respond to stockouts

The risk of a BPG stockout is reduced when:

- More brands of BPG are registered within a country. This allows different brands to be purchased during periods of shortage without regulatory or administrative delays.[467]
- Buffer stocks of BPG are held by the Government to cover periods of shortage.[467]

It may be possible for RHD control programmes to advocate for these issues.

RHD control programmes should also consider working to reduce stockouts by:

- Ensuring that BPG is identified nationally as an essential medicine. BPG is included on the WHO Model List of Essential Medicines and the Model List of Essential Medicines for Children.[468,469] Most countries also have a national essential medicines list or formulary which is not necessarily the same as the international lists. You should ensure that BPG is listed on your national essential medicines list or formulary and that RF/RHD are recorded as an appropriate Indication.
- Improve capacity for forecasting use of BPG at a national level for all indications of the product.[467] This allows for better ordering reflecting real product need. A variety of resources outline supply chain management of pharmaceuticals in low resource settings and some organisations provide technical support. WHO has collated some these resources online.

If stock outs do occur it is reasonable for governments or RHD control programmes to:

- Attempt to confirm when BPG will be available, and emphasise to the responsible authorities the critical importance of ensuring supply as soon as possible.[23]
- Attempt to identify where there are remaining supplies of BPG and decide how they should be used. It may be necessary to divert supply for treatment of syphilis in women who are pregnant.
- Communicate with healthcare staff about alternative antibiotics to be used until BPG is readily available. Clearly explain that alternative regimens are temporary and that the more effective protocol of BPG injections will be resumed when supplies are available again.[23]
- Communicate to people living with RHD about the stockout, explain attempts to secure a new supply, outline alternative secondary prophylaxis regimens and explain that they are temporary until new supplies of the product are found.

Use and administration of BPG

There are three main manufactures of raw BPG product. None of these companies have market authorisation from a 'stringent regulatory authority' for BPG. This means that production standards may vary.[467] Variation may contribute to some of the concerns about penicillin quality and safety.

> "The majority of participants expressed serious concerns regarding the availability, quality and safety of penicillin."
>
> Zühlke et al, Second RHD Forum Report, Cape Town, 2013.[290]

Pain on administration

Pain is frequently reported as a barrier to use of BPG.[466] In a New Zealand study, 405 patients (5 years of age to adult) reported a mean pain score of 5.4/10 during administration of Bicillin L-A®(Pfizer).[470] In the Middle East, 117 paediatric patients (>10 years) were given injections of powdered BPG diluted in 3.2 ml of sterile water for prophylaxis of RF. The mean score for pain on administration was 6.7/10 (range 4–10).[471] Some programmes have developed guidelines for managing fear and pain at the time of BPG injection. These are summarised in Box 19.

BOX 19:

Reducing the pain of BPG administration

A number of strategies have been developed to reduce the risk of pain when administering BPG:

- Use a 21-gauge needle – smaller needles are much more likely to block and increase pain during administration.
- Allow alcohol from swab to dry before inserting needle.
- Give the injection as soon as the solution has been mixed, blockages in the needle are more likely to form if there is a delay in administration.
- Apply pressure to the injection site with thumb for 10 seconds before inserting needle.[472]
- Deliver injection very slowly (preferably over at least 2–3 min).
- Distract patient during injection (e.g. with conversation).
- Clinical guidelines from Namibia suggest health professionals 'encourage the patient to eat before coming for the injection'.[473]

Some programmes mix BPG with a small amount of local anaesthetic to try and reduce the pain of administration.[474,475] There is good evidence that using local anaesthetic as a diluent reduces pain.[470,471,475] There is some evidence that this does not affect serum concentration of BPG.[476] However, local anaesthetic is not currently a licensed additive nor supported by BPG manufacturers.

Difficulty mixing the powdered BPG into a suspended solution

BPG can be difficult to suspend in solution, sometimes causing visible 'clumps' when the powdered product is mixed with sterile water or saline.[467,477] This is sometimes interpreted as an indication of a poor quality product. Poor mixing in water also causes blockages in the needle during delivery, potentially increasing pain during administration.

Duration of serum penicillin concentration levels

There is some research to suggest that the serum concentration of penicillin falls more quickly than expected, potentially leading to periods without enough penicillin in the blood to provide protection against Strep A infection.[465,478] The cause of this declining serum concentration is unclear. Preliminary laboratory analysis of BPG samples from 10 countries shows that all samples contained BPG within acceptable limits of potency and that there was no evidence of degradation products.[479] The implications for dose size and interval are discussed in Chapter 18.

Safety and adverse drug reactions

Communities and healthcare providers are understandably concerned about the risk of adverse drug reactions to BPG injections. In particular, there are concerns about anaphylaxis 'a severe, life-threatening, generalised or systemic hypersensitivity reaction'.[480] These fears are a major barrier to use of BPG.[467]

The best information about adverse reaction and allergy to BPG comes from a study by the International Rheumatic Fever Study Group in 1991. Between 1988 and 1990 they tracked 1790 people from 11 countries having 32,340 injections of BPG.[481] In this study 3.2% (57/1790) of people had an allergic reaction of any kind, 0.22% (4/1790) had anaphylaxis and one of the 1790 people (0.05%) died. This death represents a rate of 0.31 per 10,000 injections. This single death occurred in a 15 year old patient with severe mitral valve disease, and congestive heart failure. The rates of adverse reaction in this study were reassuringly low. Other large studies of BPG (when used for syphilis) also show the product has low rates of anaphylaxis, occurring in approximately 1 person per 100,000 administrations.[482]

Case reports of adverse reactions of BPG persist despite good data showing an acceptable risk profile for anaphylaxis.[466,483] These reports include unexplained deaths after the use of BPG. Some regulators have responded by banning the use of BPG.[466] However, it is not clear whether these deaths are caused by anaphylaxis. Other causes of collapse or unexpected death could include accidental intravenous administration or arrhythmia in people with advanced heart valve disease. Any adverse events to injection should be reported to national pharmacovigilance programmes wherever they exist. Collection of case reports may help provide more information about the contributing factors to unexpected reactions. A prospective study of adverse reactions to BPG may be needed to provide more information.

Opportunities for integration

A growing number of low and middle-income countries have pharmacovigilance programmes.[484] You may be able to work with medicines agencies to strengthen capacity for monitoring adverse drug reactions for all drugs at a local or national level. Resources are available from the WHO and partners.

Supporting BPG Administration

Strategies to support BPG administration are outlined below in Table 22.

BOX 20: Reformulation of BPG

The painful injections of BPG and the frequent dosing for secondary prophylaxis make it a difficult product to use. There have been calls for a new form of penicillin which are less painful and provide a longer duration of serum penicillin concentration.[486] This would potentially improve acceptability and outcomes from secondary prophylaxis. Active research is underway to explore this possibility.[487]

Other antibiotics

A small minority of people with RHD will have a history of penicillin allergy and be unable to receive BPG. In these cases, other oral antibiotics will be needed. A number of different oral antibiotics can be used, although all provide inferior protection from RF recurrence.[488] See summary, Chapter 18.

Benzathine penicillin G used for primary and secondary prevention of rheumatic fever.

Table 22: Strategies to support BPG administration

SUPPORTING PROVIDERS	
Supporting safe injection techniques	Safe injection techniques can reduce the risk of dangerous intravascular administration of BPG. People giving BPG should be taught about anatomical landmarks to ensure the injection is given into a large muscle mass – different sites are used in different countries. Slow and steady injection may also reduce pain and needle blockages. A locally adapted task aid for injection technique may be useful addition to local programmes.
Addressing fear of anaphylaxis	Healthcare workers should receive information about the known rates of adverse reactions and anaphylaxis to BPG and efficacy for primary and secondary prevention. In general these figures are reassuring that the risk of adverse outcomes is lower than the risk of disease progression from RHD.
Improving recognition and treatment of anaphylaxis	People administering BPG should receive detailed training about the management of anaphylaxis. The World Allergy Organization has developed guidelines and resources in different languages which may be adaptable to your setting.[480] Training and equipment will be needed for effective management of anaphylaxis. In Zambia, a large programme has been conducted to understand and address provider concerns about administering BPG, with a particular focus on primary prevention.[324] Focus group discussions identified fear of penicillin allergy as a major barrier to use. A subsequent workshop included training on identifying anaphylaxis and management of simulated patients. Flip chart resources and a kit of allergy medications were also developed to support provider confidence. A video produced during the programme is available online from the Pan Africa Society of Cardiology website . Programme participants were interviewed 4–6 months after training – 6 of 18 interviewees had now safely administered BPG injections confidently.[324]
SUPPORTING PEOPLE HAVING BPG INJECTIONS	
Education about the process of injection	In New Zealand, resources have been developed to help explain to parents and caregivers what to expect during and after a BPG injection for primary prophylaxis.[485]

18. PROVISION OF SECONDARY PROPHYLAXIS

THINGS TO CONSIDER

- Does your programme provide secondary prophylaxis?
- Do you have standard guidelines for deciding which antibiotics to use?
- How do you define and measure adherence?
- What are the major barriers to adherence in your setting?
- What strategies do you use to support adherence? Do these meet the needs of people living with RHD?
- Do you have an automated recall system for people overdue for secondary prophylaxis injections?

Once your programme has a register of people living with RHD and has established a reliable supply of antibiotics, delivery of secondary prophylaxis can begin.

Developing secondary prophylaxis guidelines

A range of guidelines for secondary prophylaxis have been developed around the world. Two international guidelines come from WHO[22] and WHF[23]. Local development and guideline adaptation has occurred in a number of other settings. Adapting or adopting these guidelines in your setting is an important first step for delivery of secondary prophylaxis. Guideline development should be overseen by a local committee of experts and address three key questions:

Antibiotic choice

Existing guidelines recommend BPG as the first line antibiotic for prophylaxis. This reflects the pharmacokinetic properties of BPG – the drug is slowly metabolised by the body, ensuring that a low penicillin level is detectable in the blood for up to four weeks after injection. The presumed mechanism of secondary prevention is for the low levels of penicillin to inhibit growth of Strep A bacteria and prevent infection should the person be exposed to the bacteria. In this way, Strep A infections and subsequent RF recurrences are reduced.

In addition to pharmacokinetic properties, Strep A is universally sensitive to penicillin and BPG is a relatively minor contributor to antibiotic resistance concerns. If administration of BPG is impossible, oral penicillin V is the usual second line choice. However, oral penicillin is less effective than BPG in preventing disease progression, even when it is taken as directed.[489]

2. Dose size and interval

Historically, protective levels of serum penicillin were thought to last 4 weeks following a standard dose of BPG [465] Some contemporary studies suggest that penicillin levels may fall below protective levels sooner, potentially within 2 weeks.[478] This has raised questions about the need to increase the dose of BPG or reduce the dose interval. There is some evidence that 2 or 3 weekly injections are more effective than 4 weekly injections.[489] However, many programmes focus on 4 weekly (28 day injection schedules) for operational reasons and to foster adherence.[347] This approach has some support – in a larger register study across a number of countries – people on 2 weekly regimens were less adherent to secondary prophylaxis than those on 3 or 4 weekly regimens.[490]

3. Duration of prophylaxis

The evidence for duration of secondary prophylaxis is complex. The risk of RF recurrence is highest following an episode of RF. Therefore, some programmes focus on delivering secondary prophylaxis for young people soon after diagnosis of RF when they at the highest risk of recurrence.[491] However, late recurrences, many years after the index event, do occur and are potentially devastating in the setting of advanced heart valve disease.[492] For this reason most clinical guidelines recommend years of secondary prophylaxis – lifelong in settings of advanced heart disease. The rationale for long term secondary prophylaxis may need ongoing reinforcement for health staff. For example, in India at least one programme has documented cases of prophylaxis being stopped after heart surgery.[493]

A summary of existing secondary prophylaxis guidelines appears in Table 23, expanded from [494]

Table 23: Summary of global secondary prophylaxis guidelines

GUIDELINE SOURCE	FIRST LINE ANTIBIOTIC	DOSE	INTERVAL	ALTERNATIVE ANTIBIOTICS	DURATION OF THERAPY	YEAR
WHO[22]	BPG	<30 kg: 0.6 U >30 kg: 1.2 U	21 days if high risk 28 days if low risk	Phenoxymethyl -penicillin 250mg twice daily	No evidence of carditis: 5 years since last attack or 18 years.* Resolved carditis: 10 years since last attack or 25 years old. Moderate-severe or surgery: lifelong.	2001
Australia[347]	BPG	BPG <20 kg: 0.6 U >20 kg: 1.2 U	4 weeks (3 weeks for selected groups)	Phenoxymethyl -penicillin 250mg twice daily	No evidence of carditis: 10 years since last attack or 21 years.* No RHD or mild: 10 years since last attack or 21 years old.* Moderate: Until 35 years old. Severe: 40 years or longer.	2012
New Zealand[375]	BPG	BPG <30 kg: 0.6 U >30 kg: 1.2 U	4 weeks (3 weeks for selected groups)	Phenoxymethyl -penicillin 250mg twice daily	None or mild RHD Minimum of 10 years after most recent episode ARF or until age 21 years (whichever is longer). Moderate RHD Until age 30 and then reassess. Severe RHD Until age 40 but reassess at age 30.	2015
India[495]	BPG	<27 kg: 0.6 U >27 kg: 1.2 U	<27 kg: 15 days >27 kg: 21 days	Phenoxymethyl -penicillin Children: 250 mg twice daily Adults: 500mg twice daily	No evidence of carditis: 5 years since last attack or 18 years.* Mild-moderate: 10 years since last attack or 25 years old. Severe RHD or post intervention: lifelong or until 40 years of age.	2008
South Africa[496]	BPG	<30 kg: 0.6–0.9 U >30 kg: 1.2 U	<30 kg: 125mg twice daily >30 kg: 250mg twice daily	Phenoxymethyl -penicillin	No evidence of carditis: 5 years since last attack or 18 years.* Resolved carditis: 10 years since last attack or 25 years old. Severe/post valve surgery: lifelong.	1997

GUIDELINE SOURCE	FIRST LINE ANTIBIOTIC	DOSE	INTERVAL	ALTERNATIVE ANTIBIOTICS	DURATION OF THERAPY	YEAR
Saudi Arabia[497]	BPG	<27 kg: 0.6 U >27 kg: 1.2 U	4 weekly	Pen V	Rheumatic fever with carditis and residual heart disease (persistent valvular disease): >10 years since last episode and at least until age 40 years, sometimes lifelong prophylaxis. Rheumatic fever with carditis but no residual heart disease (no valvular disease: For 10 years after the last attack, or at least until 21 years of age (whichever is longer). Rheumatic fever without carditis 5 years or until 21 years, whichever is longer. More severe valvular disease: Lifelong. After valve surgery: Lifelong.	2017
Sudan[386]	BPG	1.2 U	3 weekly	Not recommended	Patients without carditis: till 25 years of age. Patients with carditis: For life.	2017
Uganda[498]	Pen V 500 mg twice daily OR BPG	BPG <30 kg: 0.6 U >30 kg: 1.2 U	4 weeks	Pen V 500 mg twice daily	Rheumatic fever without carditis: for 5 years or until age 18 or 21 years old. – Carditis but no residual heart disease: for 10 years or until age 25 years old. – Carditis with residual heart disease: until age 40–45 years or for life.	2016
Namibia[473]	BPG OR Pen V 500 mg twice daily	BPG 1.2 U	4 weeks	Pen V 500 mg twice daily	Until the age of 20 years or 5 years after the attack.	2011

Supporting adherence

Adherence to the local schedule of secondary prophylaxis injections is poor worldwide. The issue is complicated by difficulties in defining, measuring and describing adherence. These issues are outlined in Box 21.

BOX 21:
Defining adherence to secondary prophylaxis

There are three main reasons to define or quantify adherence to BPG-based secondary prophylaxis regimens:

Clinical: To assess whether an individual person living with RHD is at risk of Strep A infection and RF recurrence from inadequate adherence.

Administrative: To assess how well an RHD control programme is delivering secondary prophylaxis for a population and track progress towards service delivery goals.

Research: For research activities to define the effect of adherence to BPG on disease outcomes.

The measures of adherence best suited to each of these goals differ – as does capacity to measure adherence in various settings. This had led to a wide range of variation in how adherence is reported.[499] This variation can be confusing because it makes it difficult to compare changes over time or between different locations.[500] A summary of different approaches is provided below and in Tables 24 and 25.

Table 24: Individual measures of adherence to secondary prophylaxis

MEASURE	DESCRIPTION	NOTES AND UTILITY
NUMBER OF DOSES MISSED	This is simplest approach to defining adherence – particularly for people having frequent clinical reviews. Clinicians can ask how many injections have been received since last review – 3 or 6 months earlier.	A basic measure of adherence which can be useful for individual pairs of clinician and patient. However, difficult to track over time and variable follow up schedules.
PROPORTION OF SCHEDULED INJECTIONS OVER A GIVEN PERIOD OF TIME	This measure uses the number of planned injections as a denominator. For example, people scheduled to have a dose of BPG every 28 days should have 13 injections per year. The numerator is the number of injections per year. Therefore, a patient who has had 7 injections in 1 year has an adherence rate of 7/13 = 53%. This provides an individual indication of adherence.	The proportion of scheduled injections is an intuitive measure of adherence for individual patients. There is emerging evidence that receiving 80% of scheduled injections is a biologically meaningful measure of protection at a population level.[454] However, individual patients can miss a number of consecutive injections – potentially leaving them unprotected from Strep A infections for months at a time – without that risk being clearly communicated in aggregate figures.
DAYS AT RISK	Days at risk is an evolving methodology to give a biologically meaningful measure of protection from Strep A infection. Each day overdue from a scheduled BPG injection is considered a day 'at risk'. These days are tallied throughout the year and can be presented as median days at risk.	The most complex measure of adherence. Days at risk which provides nuanced information about whether injections have been delivered on time or missed consecutively. The concept of days at risk may be helpful for communicating the importance of on time injections to individual people living with RHD.[45,501,502] However, days at risk can be difficult to calculate manually and best suited for automated electronic clinical record/register-based systems.

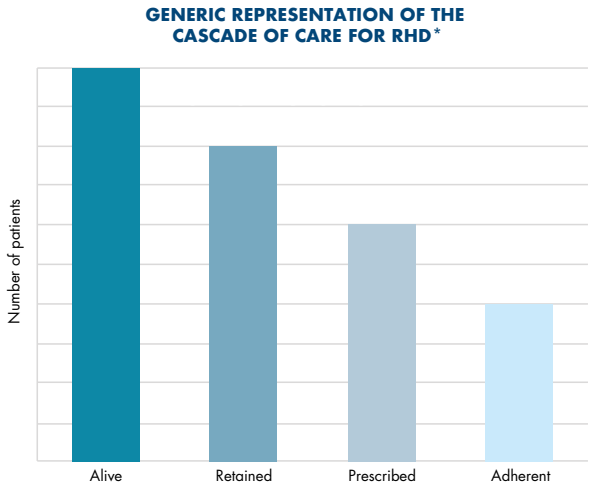
Population measures of adherence (used for administrative or research purposes) often present individual level data categorically. For example, the number of people on a register or in a clinic who are achieving 'poor' adherence or 'good' adherence.

Table 25: Population measures of adherence

MEASURE	DESCRIPTION	NOTES AND UTILITY
NUMBER OF DOSES MISSED	Missed dose data can be categorised. For example, 'poor' adherence might be defined as 0 doses over 3 months, 'reasonable' might be 1–2 doses over 3 months and 'good' adherence might be 3 doses in 3 months.	Missed doses measurements are difficult to compare between services or across time. Useful in a small number of settings.
PROPORTION OF SCHEDULED INJECTIONS	This is the most common measure of population adherence and is widely used for administration and research. For example, RHD control programmes in Australia use 'Proportion of individuals receiving > 80% of scheduled injections' as a key performance indicator.[347]	Proportion of scheduled doses can mask substantial variation between individuals in a cohort.[499] The percentage of people receiving a proportion of scheduled injection is not an intuitive measure and can be difficult to communicate to policy makers or healthcare workers as a service delivery goal.
DAYS AT RISK	Population application of days at risk may offer an opportunity for more detailed whole-of-system performance measure. Terminology for this approach is still developing, known as 'proportion of days covered' in some papers.[502]	Days at risk is biologically plausible and may communicate to policy makers and health staff the importance of timely adherence. The best measures for reporting days at risk are being developed – mean and median days at risk for individuals have been reported in a recent study.[503]

All of these population measures of adherence depend on clearly identifying who should be receiving secondary prophylaxis. This can be challenging because the denominator (the number of people who should be receiving secondary prophylaxis) changes over time. This can happen if people are lost to follow up or are no longer engaged with the health service. If these people are removed from the calculation of population adherence the proportion of people receiving sufficient prophylaxis will be artificially high.[503]

One way of addressing this problem is to think of delivery of secondary prophylaxis as part of a 'cascade of care'. This approach is adapted from measurement of care components for HIV.[454] The cascade of care is based on the number of people alive, the number retained in care (receiving active follow up), the number of people prescribed secondary prophylaxis and the number 'adherent' to secondary prophylaxis (defined using one of the measures outlined above). This approach is exemplified in a 2017 publication from Uganda which identified that people living with RHD engaged in clinical follow up were also adherent secondary prophylaxis. The main determinant of poor secondary prophylaxis adherence occurred when people were no longer retained in active clinical care.[454] This information would be lost in a standard analysis of percent of people receiving a proportion of scheduled injections' analysis. Distinguishing between people who are poorly adherent from those who have been lost to follow up entirely may offer new ways to support care delivery.



*Illustrative diagram only, not real data.

Figure 14: Determinants of adherence

BOX 22:

Language used to discuss secondary prophylaxis adherence

Internationally, the term 'compliance' is often used to describe whether or not individuals prescribed medication are taking that medication. However, the term can also be interpreted as whether people living with RHD are 'doing what they're told'. Given the challenges in affording, accessing and adhering to secondary prophylaxis it may be unwise to attribute all the responsibility for taking medications to individuals.

Using language which reflects shared responsibility for health may help reduce stigma and frustration with individuals perceived to be 'non-compliant'. Sometimes this is called concordance, to reflect agreement to the treatment plan between the person receiving care and the healthcare worker.[505] In this handbook we have used the term 'adherence' to refer to delivery of scheduled medication.

"Within this context the term 'poor compliance', often used by health professionals and administrators, would best be replaced by 'poor service' in the majority of circumstances."

McDonald et al, Outcomes of cardiac surgery in Aboriginal Australians, 2004.[504]

Contributions to poor adherence and opportunities to improve

There are many individual, systematic and social factors which influence whether people living with RHD will receive recommended secondary prophylaxis injections of BPG (see Table 26).[506,507]

A number of studies to explore determinants of adherence have been conducted around the world – sometimes these studies find common factors and others have differing results. The specific determinants of adherence in different locations and communities are likely to vary. One way to consider and address determinants of adherence for your programme is to use the 'determinants of adherence' framework developed by the WHO.[508] This model is reproduced In Figure 15 and outlines the 5 different domains which influence medication adherence.

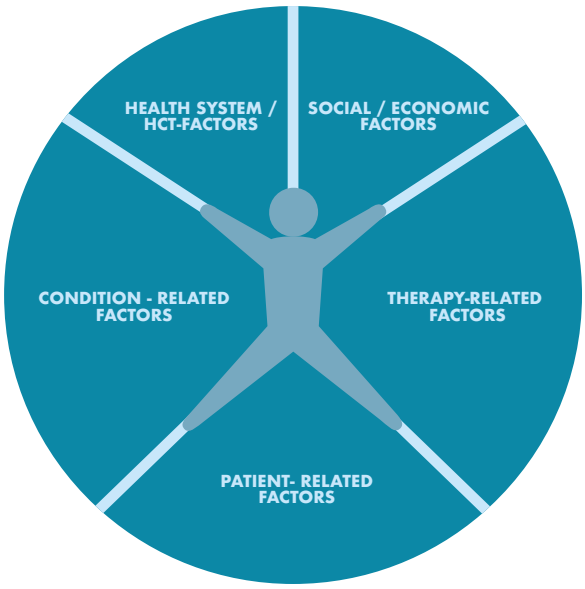


Figure 15: Determinants of adherence to long-term therapies.[508]

Table 26: Contributors to adherence and strategies to address low adherence to secondary prophylaxis injections

CONTRIBUTOR TO ADHERENCE	STRATEGIES TO IMPROVE ADHERENCE
CONDITION RELATED FACTORS	
PERCEPTION AND UNDERSTANDING OF ILLNESS AND MEDICATION One of the greatest challenges in delivery of secondary prophylaxis is that the people who need it to prevent disease progression generally feel well. Young people and their families may be understandably doubtful or confused about the importance of secondary prophylaxis when they experience no obvious signs of disease. For example, 'doubts about the need for prophylaxis' were a barrier to compliance in a small study in Mumbai.[509] In New Caledonia, young people who had experienced a symptomatic episode of RF were more likely to be adherent to secondary prophylaxis than those who had always been asymptomatic.[510] See further discussion of adherence following echocardiographic-screen diagnosis in Chapter 20]. Cultural beliefs, including the role of traditional medicines may also contribute to adherence behaviours.	**CONSISTENT MESSAGES FROM HEALTHCARE PROVIDERS** "Every contact with a health professional that does not discuss secondary prophylaxis is a substandard consultation." Wilson, Secondary prophylaxis for rheumatic fever: simple concepts, difficult delivery, 2013.[500] Consistent messages about the value of secondary prophylaxis should be provided to all healthcare staff, communities and people living with RHD. In Jamaica, over 80% of children and adults living with RHD agreed that encouragement from a doctor or nurse was one of the reasons they took their injections.[511] All staff should be empowered to discuss adherence. 100% of scheduled injections should be the goal for every person receiving secondary prophylaxis.[500]
HEALTH SYSTEM FACTORS	
EXPERIENCE OF INJECTION ADMINISTRATION The experiences that people have when receiving injections can be an important determinant of whether they will keep returning for injections. Qualitative studies suggest that supportive relationships with clinical staff encourage trust and support return attendance.[224,459] In particular, some people receiving BPG injections report that having a good relationship with the health worker providing injections helps makes the interaction more predictable and less intimidating.[512,513]	**EMPLOY OR IDENTIFY REGULAR STAFF TO DELIVER SECONDARY PROPHYLAXIS** Clinics may be able to facilitate supportive relationships by nominating a single dedicated healthcare worker responsible for administering BPG injections. Some programmes focus on building relationships when people begin secondary prophylaxis and providing supportive education about the need for ongoing treatment.[514] Evidence from Australia suggests that this approach can improve adherence.[317,459,515] Similarly, in Brazil the 'combined actions of a multidisciplinary health team in providing information and improving the clinical assistance and relationships with patients and their families' has been identified as key elements to support improved adherence.[48]

CONTRIBUTOR TO ADHERENCE	STRATEGIES TO IMPROVE ADHERENCE
EXPERIENCE OF HEALTH SYSTEM INTERACTIONS People living with RHD and receiving regular secondary prophylaxis injections are in frequent contact with the health system. This can mean that administrative or logistical frustrations of care are amplified on each visit. Long wait times for injection administration is frequently identified as a barrier to adherence.[511]	**FAST TRACK OR DEDICATED APPOINTMENT TIMES** It may be possible to reduce waiting times by allowing people waiting for BPG injections to be prioritised for treatment. This may be by using a 'fast track' card or other signal. Alternatively, some clinics administer all secondary prophylaxis injections in a dedicated clinic ensuring that people can pass more speedily through the process.
GEOGRAPHIC LOCATION OF INJECTION ADMINISTRATION In some places people must travel long distances to receive secondary prophylaxis injections. In Jamaica 20–30% of surveyed people living with RHD reported long travel distances as a barrier to adherence.[511] In Thailand in the 1980s approximately 200 people receiving monthly BPG injections were randomised to centralised dispensing (main hospital paediatric department) or decentralised dispensing (local health centres with 3 monthly specialised review). On average, people in the decentralised group received ten injections annually and people returning to the central hospital each month received only five injections.[516]	**DECENTRALISED DISPENSING AND ADMINISTRATION** Local health staff may be able to source, prescribe, dispense or administer secondary prophylaxis in their communities, rather than rely entirely on central providers in larger centres. This may be 'convenient as far as time and travel expense are concerned'.[517,518] For example, in Kiribati, the RHD control programme worked with the Ministry of Health to shift delivery of BPG from the central hospital to supported local clinics.[273] Legal and regulatory systems may be needed to make this possible – particularly if injections are to be provided by health workers or other people who do not usually administer injections.[519,520] **MOBILE INJECTION DELIVERY** Some programmes can fund home visits by nurses or care workers to deliver secondary prophylaxis injections. Although potentially expensive, this approach reduces inconvenience for people living with RHD and maximises the opportunities for adherence.[343,512] In some part of Australia and New Zealand this approach has been associated with high rates of adherence.[343,512] Programmes considering mobile injection delivery should address the safety of healthcare workers (including risks from pets, violence and travel) as well as their capacity to manage adverse drug reactions in the community Home visits may also provide an opportunity to evaluate living conditions, and provide education to family groups.
SOCIOECONOMIC FACTORS	
COST AND INCONVENIENCE Financial cost is an important determinant of adherence with secondary prophylaxis. In Egypt, children who had to pay for BPG were much less likely to receive the recommended number of doses each year.[521] In some other places people must take time off work or school to travel for injections causing indirect costs in time and travel. Cost of injections, costs of travel and disruption are widely identified as barriers to adherence.[522]	**ENSURING AFFORDABLE BPG FOR PEOPLE LIVING WITH RHD** A number of successful RHD programmes have secured a supply of BPG which is free to some, or to all, people receiving secondary prophylaxis.[327,461]

CONTRIBUTOR TO ADHERENCE	STRATEGIES TO IMPROVE ADHERENCE
THERAPY RELATED FACTORS	
REMEMBERING TO HAVE INJECTIONS Remembering to have injections, particularly when they only occur every few weeks, can be challenging. This may be exacerbated in low income families or when large family size means there are many children with different health issues.	PRODUCE PROPHYLAXIS CARDS People living with RHD can be given a card which can be used to record BPG administration dates. The cards may help people living with RHD remember when their next injection is due and can also provide clinicians an indication of whether injections have been missed or given elsewhere. Development of prophylaxis cards and monitoring of adherence was associated with improved prophylaxis in an historic programme in Barbados.[523] In Brazil and Nepal cards are also used as a reminder for upcoming injections and to monitor adherence.[48,327] In Uganda, BPG administration cards are also use to measure medication adherence for research outcomes.[454] MEMORY CUES AND REMINDERS **Text messaging and phone calls** Text messages (SMS or texting) can be used to remind people that injections are due though this approach hasn't been evaluated.[453] In the Pacific Islands phone calls or long distance radio messages have been used to encourage people to return for secondary prophylaxis delivery.[303] **Calendars** RHD reminder calendars have been developed in South Australia and in Fiji.[524] These are distributed to people living with RHD to provide information about the condition, positive messages about healthy choices and to provide a regular reminder about BPG injections. **Apps and other electronic reminders** In Australia two mobile phone applications have been developed to support adherence with BPG injections.[525,526] Both can be downloaded from the app store and customised to individual treatment regimens. Evaluation of apps for treatment support in the Australian Indigenous setting are underway. A Facebook app has also been developed to provide an alternative reminder system which can be used across multiple mobile devices.[527] **Full moon reminders** In 2006, the Central Australia RHD control programme launched a novel effort to encourage people with RHD to time BPG injections with the full moon.[528] The 'full moon strategy' was developed to reflect traditional approaches of Indigenous people living with RHD. A range of interventions – including personal calendar cards, full moon posters and radio advertisements – were developed. A moderate increase in BPG uptake was demonstrated with more consistent uptake during the full moon. [528]
FEAR OF INJECTION PAIN Pain from intramuscular injections of BPG is likely to be a barrier to adherence, though individual experience is mixed.[529] In Jamaica, 78% of children reported missing their injections because they feared the painful experience.[511] In Uganda, fear of injection pain was also commonly identified as a reason for missing injections.[530]	MINIMISE PAIN FROM INJECTIONS Some injection protocols or guidelines have been developed to reduce the pain associated with BPG injection – these are outlined in Chapter 17.[531] A detailed task aid has been developed in New Zealand for nurses to use when giving BPG injections.[474] PROVIDING INCENTIVES FOR HAVING INJECTIONS Some reviews of BPG adherence have suggested that people living with RHD could receive an incentive to encourage adherence with secondary prophylaxis injections.[506,532] Small rewards for injection delivery have been used as informal incentives but not yet evaluated.[533]

CONTRIBUTOR TO ADHERENCE	STRATEGIES TO IMPROVE ADHERENCE
FEAR OF ALLERGY OR ADVERSE REACTION In some places people living with RHD and/or their health workers fear adverse reactions to BPG injections. This may be a fear of allergy, anaphylaxis or sudden death.[534]	**PROVIDE INFORMATION ABOUT INJECTIONS** In general, BPG injections are safe and effective. However, some adverse events have been reported and this can be very worrying for people who need ongoing injections. Adverse reactions to BPG are discussed in more detail in Chapter 17.
STOCKOUTS OF BPG Shortages of BPG contribute to poor adherence in settings with unstable antibiotic supply.[507]	Issues with BPG supply are addressed in Chapter 17.
CONDITION RELATED FACTORS	
METHOD OF DIAGNOSIS There may be a difference in secondary prophylaxis adherence between people who have had an episode of symptomatic RF and those who have been identified as having RHD on echocardiographic screening. It is possible that people who have never felt unwell with RF, or have had less inpatient education about RHD, may not be as adherent with ongoing BPG injections.[502] This association is likely to vary by setting.[503]	**PROVIDE EDUCATION** It is reasonable to ensure that people diagnosed through echocardiographic screening have intensive education soon after the echocardiogram as a foundation for ongoing adherence. This should be part of obtaining informed consent for echocardiographic screening and is discussed further in Chapter 20.
PATIENT RELATED FACTORS	
AGE Delivery of secondary prophylaxis in early childhood years is often facilitated by parents or caregivers. A number of studies demonstrate a drop off in adherence as adolescents begin to take responsibility for their own prophylaxis adherence.[502,503]	**TRANSITION CARE** Focused strategies and transition care from paediatric to adult services may be an appropriate strategy to address this issue.[529] **PEER SUPPORT** In one informal programme in Australia, clinic staff facilitated young people with RHD to provide 'mental and emotional support' to each other while receiving their RHD injection.[535] The role of peer support for prophylaxis adherence requires further investigation.

"The key to better compliance was enthusiastic, dedicated staff in the Rheumatic Fever Programme and the reminder phone call to remind the patients of the injections."

Viali et al, Rheumatic fever programme in Samoa, 2011.[453]

Strategies to improve secondary prophylaxis adherence are numerous, and often a combination of strategies that suit the needs of the patient, healthcare provider and setting, are required for better impact. Appropriateness of chosen strategies differs across settings and should be considered locally and collaboratively.

19. PRIORITY-BASED FOLLOW UP

THINGS TO CONSIDER

- How will you follow up people with RHD who need specialist input?
- How will you ensure the people with the greatest need receive the greatest care?
- How will priority based guidelines and protocols be disseminated throughout the region?
- How will clinicians be informed about their use?

Developing an RHD register helps to improve delivery of secondary prophylaxis. A register can also facilitate a comprehensive follow up programme for people living with RHD. In this case, the register will include people living with RHD at very different stages of disease. Some people on the register will have a history of RF and be on prophylaxis to prevent recurrences, some will have no symptoms, others will have severe disease, advanced heart failure, be awaiting surgery or needing post-operative follow up. People with RHD who are deceased should also captured on the register for calculating mortality rates. The clinical needs of these patients vary and a system is needed to ensure that patients who need the most input are provided with the most support.

Assigning priorities to different groups of patients is one way of approaching this problem. A 'priority-based follow up' system provides a framework for scheduling and arranging follow up.[10,23,47,71] For example, RHD may be catagorised as mild, moderate or severe, which allows frequency of clinical follow up to be appropriately assigned. This priority-based system is similar to clinical staging systems used to describe and triage interventions for other diseases. In developing countries these may include the stages of chronic kidney disease and severity stages of chronic obstructive pulmonary disease.[536,537]

The benefits of a priority-based approach are outlined in Table 27.

Table 27: Benefits and precautions of priority-based RHD care

BENEFITS OF A PRIORITY SYSTEM	PRECAUTIONS OF A PRIORITY SYSTEM
• Helps ensure that the most resource intensive care is targeted to the people who need it most. • Provides local health staff with a consistent framework for managing clinical issues. • Useful in locations where staff have limited experience or training for managing people with RF and RHD or staff turnover is high. • Supports self-management of RHD by people living with disease. • Data on disease severity and progression can be used for epidemiologic and programme evaluation purposes.	• Expert clinicians need to agree on the categories and the criteria, or confusion may arise. • Primary health clinicians need to be able to access information, education and training regarding the priority system. • Resources and services that are recommended within the priority system need to be accessible, or primary health clinics won't be able to fulfill the care planning activities. • Specialist clinicians need to support and act as role models to demonstrate the use of priority-based guidelines to support application by all staff.

Steps to develop a priority-based system

A priority-based system will need to be developed by local clinicians and experts to reflect feasibility and available resources. There are four main stages of development:

1. Establishing categories of disease severity

An initial step in developing priority categories is gaining consensus on the categories of heart disease severity. A number of categories already exist, including the New York Heart Association (NYHA) classification of symptomatic heart failure and the RHD Australia 'priority-based' categories for RHD follow up, reproduced in Table 28.

Priority categories for follow up may need to be adapted for your setting to reflect access to echocardiography and/or surgical services. For example, it may be more appropriate for priority categories to be based on clinical signs and symptoms rather than echocardiographic classification. Patients on anticoagulation or planning pregnancy may also need special consideration.

2. Establishing follow up recommendations

In settings where a primary health system is established, follow up recommendations should be developed in consultation with doctors in health clinics. Ideally, follow up activities are integrated into the activities of primary health workers, with RHD control programme workers providing resources, education and support.

However, in some settings follow up activities will be undertaken by clinicians in the tertiary sector and/or in partnership between both tertiary and primary sectors. The aim is to provide clinical care and follow up activities in line with best practice and based in evidence that is applicable to the setting.

In some places, detailed clinical guidelines have been summarised to highlight critical priorities for front line health staff.[538]

3. Develop standardised care plans

Disease categories and follow up recommendations are ideally developed into a 'care plan' which outlines the expected pathway for follow up and indications to increase or decrease the level of care.[46] Plans should be integrated and recorded within the patient information and recall system, and the local healthcare record.

4. Develop individualised care plans

Some people will need an individualised approach, including people with advanced heart disease or women with RHD planning pregnancy. Ideally, specialist clinicians determine the course of treatment, follow up plans and other management details for these individuals. Where resources permit, individualised care plans for all patients may be possible.

Opportunities for integration

An integrated care plan will take into account other aspects of an individual's medical and psychosocial needs where possible. This is particularly important when the patient has co-morbidities (other illnesses) or needs routine clinical care such as growth measurement, family planning, counselling or vaccination.

Table 28: RHD Priority Classifications adapted from RHD Australia guidelines[46]

PRIORITY 1	Severe valvular disease on echocardiography OR Symptomatic valvular disease OR Post-operative patients (mechanical prosthetic valves, tissues prosthetic valves and valve repairs including balloon valuloplasty).
PRIORITY 2	Any moderate valve disease on echocardiography, normal left ventricular function and no symptoms.
PRIORITY 3	History of RF with no evidence of RHD on echocardiography OR Trivial/mild heart valve disease on echocardiography.
PRIORITY 4	History of RF with no evidence of RHD when secondary prophylaxis has ceased.

Implementing priority-based follow up

Providing clinical care and developing follow up categories may be a novel approach in some settings. Support to deliver care this way is needed throughout the health system and it can take time for the benefits to become apparent.

Communicate priority category

The benefits of priority-based care are only possible if everyone involved in clinical service delivery is aware of the priority category of each patient. This information can be recorded in the RHD register and documented in specialist clinical letters. Recording and disseminating priority categories can take time to become standard practice. For example, a review of register details for people diagnosed with RHD between 1999 and 2012 in the Northern Territory of Australia revealed that only 73% of people had priority data recorded on the register within a year of diagnosis.[539]

Supporting self-management

Care plans are widely used in primary care in high resource settings to inform management of chronic health conditions. The process, implementation and documentation of care plans varies widely.[540] Many focus on supporting people living with disease to take an active role in self management or develop plans in conjunction with their clinical team. This may also be applicable to management of RHD. Developing written RHD plans with people living with the disease may facilitate improved care delivery.[541] In some places it may be possible to give people living with RHD a copy of their own agreed care plan as part of information sharing.

Strengthening referral systems

Referrals between primary, secondary, tertiary and quaternary health services are a risky period for loss of clinical information or breakdown in continuity of care.[459] (See Table 1). The transition from primary to secondary and tertiary care can be overwhelming for patients who may need to travel to large cities, be assessed in unfamiliar languages and be separated from community support structures.[47,459] Transport may be difficult or prohibitively expensive.[522,542] Echocardiography and specialist clinical review is often limited in low and middle-income settings, necessitating long waiting periods and further opportunities for people to be lost to follow up. Patients may be 'lost' during the referral process, particularly in settings without clearly established pathways for escalating care. These problems can be even more severe when patients are being referred or treated internationally (see Chapter 25).

> "During the first three years of the Pacific Island Health Care Project referrals were made by letter, long distance telephone or fax. Diplomatic pouch was used on several occasions... Referrals by long distance telephone calls, letters, or fax were hard to understand, often interrupted, dropped, garbled or otherwise unintelligible."
>
> Abbas et al, The Pacific Islands Health Care Project, 2008.[303]

20. ACTIVE CASE FINDING (ECHOCARDIOGRAPHIC SCREENING)

THINGS TO CONSIDER

- Are you able to deliver high quality secondary prophylaxis to people already on your register?
- How would you provide follow up for people with RHD identified through screening?
- What are the local standards of consent for screening procedures?

"There are very good examples of success stories with rheumatic control that do not involve the use of echo screening. Implementation of penicillin prophylaxis is the single biggest challenge in rheumatic heart disease prevention."

Chowdhury, The 2017 Seventh World Congress of Pediatric Cardiology and Cardiac Surgery; week in review - ambulatory pediatric cardiology.[543]

An introduction to screening

Health screening programmes are designed to 'discover those among the apparently well who are in fact suffering from the disease'.[544] Screening is a specialised issue in medicine and public health because it involves actively seeking disease in people who would otherwise be considered well. This proactive approach raises unique practical and ethical issues.

WHO have supported auscultation (stethoscope) screening of children for RHD in high risk populations since the 1970s.[545] A WHO supported auscultation screening programme began in 1984, and included 1.4 million school children in 16 countries.[546] In 2001 the WHO Expert Committee for RF and RHD again recommended auscultation screening for high risk populations.[441] Since then the advent of increasingly portable, low cost echocardiography (echo) has revolutionised screening for RHD, offering new risks and benefits.

The role of echocardiography in screening for RHD began to be explored in the mid-1990s. In 2007 the landmark paper 'Prevalence of RHD Detected by Echocardiographic Screening' was published by Marijon and colleagues, confirming a significantly increased prevalence of RHD on echocardiographic screening compared with auscultation screening.[547] Since then a vast number of echocardiography screening projects have been undertaken around the world.[548]

The vast majority of echocardiographic screening projects to date have been conducted as research activities to provide baseline descriptive epidemiology and burden of disease data. There is an ongoing debate about whether echocardiographic screening for clinician purposes (to change outcomes for people living with RHD) is a feasible possibility.[549]

Many countries have criteria to establish when population screening is appropriate, and what issues need to be considered. One of the well-known criteria are presented in Table 29.[544] The appropriateness of echocardiography screening for RHD has been assessed against these critiera.[550,551]

One of the challenges in assessing the role of echocardiographic screening in the management of RHD has been to identify what is a normal heart valve appearance and for asymptomatic young people. Defining 'screen positive' disease has been challenging. There are many small variations of normal heart valves, and it may be difficult to distinguish normal valves from early heart valve changes in RHD. Early studies of echocardiographic screening for RHD all used slightly different criteria to define RHD, making it difficult to interpret and compare results from around the world.[555] This variation in diagnostic criteria result in large variations in burden of disease and were caused by subtle changes in criteria for diagnosis.[550]

In 2012 a group of experts developed a rigorous approach to diagnose subclinical RHD: the WHF criteria for echocardiographic diagnosis of rheumatic heart disease.[64] These criteria define the morphology (shape) and functional changes of valves affected by RHD.

Table 29: Suitability of RHD for echocardiographic screening

Evidence of a significant burden of disease	There is a significant burden of RHD in low and middle income countries and in some vulnerable populations in high income countries.[2] In settings with the highest burden of disease most people present with advanced heart valve damage at the time of diagnosis and had a high rate of premature death.[5,490] The impact of RHD is particularly significant for women in pregnancy.[552]
Condition must have a latent stage	RHD has an asymptomatic phase when heart valve damage can be detected by auscultation or echocardiography but before symptoms are evident.[64] This asymptomatic phase is thought to represent carditis during episodes of RF which were not detected. About 40% of people who present with symptomatic RHD have no record of any episodes of RF.[64]
The latent stage must be detectable by simple, accessible and sensitive tests	Echocardiography is a relatively non-invasive procedure with painless application of the scanning probe to the chest wall.[550] In general, the echocardiography screening procedure appears acceptable in communities where it has been used.[553] The sensitivity of echocardiography for RHD screening depends on the screening protocol used, the screening device used, the operator and the population screened. At least some of these combinations produce acceptable sensitivity compared with expert-operator diagnostic echocardiography with standard machines. The introduction of lower cost and hand held echocardiography machines has made screening more accessible but the association with sensitivity remains the subject of ongoing research.
The early stage of disease must be treatable with adequate therapy	Regular secondary prophylaxis prevents RF recurrences and minimises progression in valve lesions in clinically diagnosed RF and RHD. It is not entirely clear that this can be directly extrapolated to asymptomatic disease diagnosed on echocardiography. Delivery of high quality prophylaxis is difficult in many parts of the world. For example, in Fiji in between 2011 and 2014 only 58.9% of people diagnosed with RHD on echocardiographic screening ever received a dose of BPG for secondary prophylaxis.[502] Therefore echocardiographic screening for RHD may not be indicated when adequate therapy is not available to alter disease outcomes. "However, mass screening for RHD and other chronic health conditions is not a viable option in most country settings, and certainly not in South Africa nor Ethiopia–both because such screening is resource-intense and unaffordable, but also because a consistent referral service response, as a minimum requirement of a good and ethical screening programme, is not available." Engel et al, Prevalence of rheumatic heart disease in 4720 asymptomatic scholars from South Africa and Ethiopia, 2015.[554]
Early intervention must improve prognosis	The natural history of subclinical RHD has not been fully established, and the progression, stability or regression of valve lesions remains unclear.

Potential benefits of echocardiographic screening projects

Potential benefits for individuals

Acceptability and education opportunities: Discussing screening projects and securing informed consent for echocardiographic screening can provide novel opportunities for large scale education about RHD. For example, in Brazil 29,695 children were exposed to an RHD education curriculum as part of a screening programme.[312] In New Zealand, an echocardiographic screening programme may have had some impact on the participant understanding of RHD and subsequent sore throat health seeking behavior.[556]

In addition, echocardiography is a relatively accessible and intuitive form of medical imaging which may foster engagement of individual and communities in heart health and health literacy.

Most studies on the acceptability of echocardiographic screening for RHD have demonstrated that participants, families and teachers feel very positive about participating in screening.[553,557] However, this may be different in other settings or depend on pre-screen education, privacy during screening or follow up outcomes. Popularity of screening programmes is common and does not necessarily reflect clinical benefit to individuals or communities.[556]

Potential benefits to the health system

Potential for high quality burden of disease data: Local, measured, high quality burden of disease data is a powerful motivator for decision makers to focus on RHD control. This is important for securing programme funding (Chapter 3), government engagement (Chapter 6), vaccine advocacy (Chapter 15) and many elements of service delivery planning. As discussed in Chapter 1, echocardiographic screening studies provide the best evidence of RHD in a given population. Therefore echocardiography screening studies may potentiate evidence-based prioritisation of RHD.

Increased awareness and engagement towards RHD: Echocardiographic screening projects may be associated with positive externalities – particularly increased awareness and attention to RHD control.[558] This has been an explicit focus of some screening activities.[169] Political action on RHD, including the Addis Ababa Communique, has been partially attributed to the influence of the many screening activities in school children in Africa.[559]

> "To date, perhaps the greatest impact of echocardiography screening programmes for RHD has been to stimulate interest in, and advocacy for, RHD control."
>
> World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease.[64]

Improvements in technology and telehealth: Echocardiographic screening projects can galvanise improvements in telehealth and other technologies. For example, in Brazil the addition of an RHD screening project provided new momentum to an existing Telehealth Network of Minas Gerais.[312] In particular, cloud-based storage of echocardiographic images has facilitated new relationships between collaborators in countries with both high and low burden of disease.[560]

Research capacity and collaboration: Increasing access to echocardiographic screening in RHD endemic, low resource settings has provided new opportunities for scientific collaboration and partnerships.[561]

Potential risks of echocardiographic screening projects

Harms to the people being screened

Diagnosis without access to secondary prophylaxis: In order for echocardiographic screening to be clinically useful it must be possible to deliver a treatment to change the course of the disease. Therefore, screening must also be coupled with a robust programme with demonstrated capacity to deliver secondary prophylaxis. This capacity does not exist in many places with a high burden of RHD.[559]

> "Therefore it follows that it is unethical to begin a screening programme if secondary penicillin delivery is not available in the region being screened."[562,563]

Diagnosis with overtreatment: Some echocardiographic screening protocols have reduced specificity for RHD which means more people will be 'false positive' on echocardiographic screening. They will be identified as potentially having RHD when in fact they do not have the disease. Unless the echocardiographic screening result is reviewed by an experienced clinician these children may receive painful monthly injections without gaining any benefit from the procedure.

Quality of life impact: The process of undergoing echocardiographic screening may be associated with reduced quality of life – particularly in children who are identified as echocardiographic screen positive and their parents.[564] This may result in changes to how children are treated by their parents – changes which can be health promoting or health harming.[557] For example, in New Zealand children with RHD detected during echocardiographic screening reduced their physical activity after the screening event.[407] In Uganda, children who screened positive for RHD during echocardiography had reduced physical and emotional quality of life.[565] This impact could be mitigated by peer support groups.[566]

Harms to the health system

Opportunity cost of resources: The financial and human resources devoted to echocardiographic screening generally reduce capacity to deliver other components of RHD control programmes. For example, expert cardiologists may have to spend their time reviewing screening images rather than scheduled follow up clinics for people already known to be living with RHD. In resource-limited settings with fragile referral systems and very little access to specialist paediatric cardiologists for follow up of RHD these constraints are often prohibitive.[559]

Unknowns about echocardiographic screening projects

The best approach to different elements of echocardiographic screening projects are unknown.[549] 'Questions still remain to be answered before we can advocate for echocardiography-based screening for RHD as an effective means of RHD prevention.'[567]

Does screening for early detection of RHD change clinical outcomes? Technical capacity for echocardiographic screening in RHD has only been available for decades. Therefore, details of the natural history of subclinical RHD (detectable only on echocardiography) is unclear. It is likely that some early heart valve lesions improve (regress), some are stable and other heart valve lesions progress to clinically significant RHD. The risk and rate of this progression is poorly understood.[568] The WHF criteria for echocardiographic diagnosis define two categories of subclinical RHD: borderline RHD and definite RHD. There is general consensus that asymptomatic people with definite RHD on screening echocardiogram are at risk of valve disease progression and should receive secondary prophylaxis.[569] However, the role of secondary prophylaxis in borderline disease is unclear and results from follow up studies are variable.[568,570] More studies are needed to know whether secondary prophylaxis for borderline RHD changes echocardiographic or clinical outcomes. These studies will take time because long term follow up is required.[571]

Which populations should be screened? The choice of population for echocardiographic screening studies influences the capacity of people to benefit from being screened and therefore how ethical it is to undertake screening.

Age: The prevalence of RHD rises with age, therefore screening adults will detect more people with RHD than screening children. However, RHD is more likely to be severe in older people who have less capacity to benefit from secondary prophylaxis and potentially undermines the goal of early diagnosis through screening.[559] It may be that repeat or serial screenings are needed to address some of these issues but the screening interval and feasibility have not been established.[569,572]

School cohorts: Screening of school children is convenient because of the large number of young people from the target age group being in a single location. However, in settings with poor school attendance, children at greatest risk of RHD may be the ones least likely to be attending school. This raises issues of both inequity of accessing care and inaccuracy in burden of disease estimates. Lack of parental presence while children are at school and difficulties securing parental consent for screening may further complicate school-based programmes.[554,573] A survey of teachers involved in a school screening project in Uganda also reflected impacts on the school, including disruption and 'too many people present'.[553] In Brazil a new model of screening through primary care clinics may offer an opportunity to improve screening coverage.[554]

What screening criteria should be used? The WHF guidelines on echocardiographic screening are a detailed technical resource with a high degree of precision. They require considerable training to use and take time to acquire all the necessary images during screening.[559,569] Therefore, simplified echocardiographic screening protocols have been used in a number of large scale screening projects. These simplified echocardiography protocols have a trade off in sensitivity and specificity.[559] There is no clear consensus about the best modifications to the WHF criteria.[570]

What kind of echocardiography machine should be used? A range of ultrasound machines with echocardiography capacity are available. Standard portable echocardiography (STAND) machines are expensive for low resource settings, may be difficult to transport and generally require mains power to function.[569] However, they provide good quality images which are required to use some of the protocols for RHD diagnosis. Handheld echocardiography machines (HAND) are less expensive, smaller and can be battery powered.[569] However, some technical parameters of HAND devices are different to STAND and impact the sensitivity and specificity of the screening.[569,570] In particular, HAND do not currently have capacity for spectral Doppler required to apply the WHF criteria for the diagnosis of RHD.[558] Battery life may be short and some issues with overheating and entry of patient details have been described.[569,570] A systematic review of STAND and HAND capacities is planned but no clear best practice approach has emerged to date.[574]

How should the diagnosis of RHD be made? Most existing protocols for echocardiographic screening rely on a 'two-step' model, a short screening echocardiogram followed by a detailed diagnostic echocardiogram by an expert operator for people who are 'screen positive'. Where necessary, education and secondary prophylaxis are usually initiated after the detailed confirmatory issue. Expert human resource constraints mean this model is not practical at scale in low resource settings. The thresholds for making a formal diagnosis of RHD by non-expert operators or using limited view protocols are not yet clear.[569]

Who should do the screening? Human resource constraints in the low resource settings with a high burden of RHD mean that screening by clinical experts (cardiology staff or specialist cardiac echocardiographs) is rarely possible. Population echocardiographic screening studies can not clearly be sustained or scaled up if dependent on visiting clinical experts. This has prompted a growing number of projects to explore whether non-expert operators can be trained to screen for RHD on echocardiography. Programmes training nurses, medical officers and medical students have been described with a varied range of training time, resources and assessment.[309,310,575] In general, it seems most non-expert operations can become competent at simplified protocols for screening echocardiography with effective support and supervision.[309,576] However, the optimal model of training, screening protocol and quality assessment is not yet clear.[549,559]

Is echocardiographic screening cost effective? It is difficult to evaluate the cost effectiveness of echocardiographic screening while there are currently so many variables about how screening could be delivered, in which populations and what kind of capacity for secondary prophylaxis delivery is required. Early economic analysis of RHD echocardiographic screening suggested that the screening could be cost effective based on historic estimates of disease progression.[105,106] A detailed analysis from Australia grounded in real data shows screening can be cost effective in this setting.[577] However, application in lower resource settings with different costs and variables remains uncertain.

"We emphasise the importance of having a well-run and effective secondary prophylaxis programme in place before embarking on larger-scale screening, and the capacity to upscale this programme with newly detected patients."

Remenyi, et al, The World Heart Federation Criteria for Echocardiographic Diagnosis of RHD 2013.[64]

BOX 23:
Initiating echocardiographic screening studies

The technical specifications of contemporary echocardiographic screening studies are increasingly complex with evolutions in protocols, echocardiography machines and providers and target populations. These scientific details are outside the scope of TIPs. Individuals and programmes considering echocardiographic screening should seek expert advice at the time of project planning. Protocols for new studies should harmonise with global best practice and collect data which can be a pool to strengthen future analysis.[578]

BOX 24:
Screening for RHD by auscultation

Screening for RHD by auscultation alone is no longer appropriate.[550] Contemporary auscultation studies consistently demonstrate unacceptably low sensitivity and positive predictive value.[579,580]

Dr Bo Remenyi and Prof Jonathan Carapetis review echocardiography during an RHD screening project in Fiji.

TERTIARY INTERVENTIONS

Historically, RF and RHD control programmes have been predominantly concerned with primary and secondary interventions. Opportunities for tertiary interventions have typically been delivered by cardiothoracic surgeons, philanthropic groups or medical missions. In some settings this has occurred in isolation from primordial, primary and secondary approaches. However, the high cost of surgical intervention for RHD has been a motivator for governments in some countries to strengthen the development of comprehensive programmes. Making an explicit link between the treatment of RHD – and the opportunities for prevention – is critical for the development of a truly comprehensive programme.

Advanced medical care is often the most urgent and obvious need when RHD programmes begin. Globally, most people present with advanced heart disease when they are first diagnosed. For example, in Uganda, 46% of new patients at a tertiary cardiac centre with a definite diagnosis of RHD had heart failure and 20% were in atrial fibrillation.[444] In a registry study of 1332 patients in 7 countries across Central and West Africa, 83% of people with RHD required surgical intervention. Only 2.2% of those people were able to receive the required intervention.[6] The widespread need for advanced medical and surgical care is not reflected in service availability.

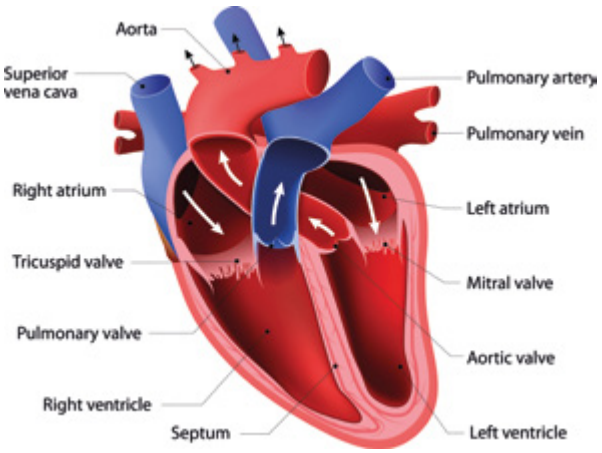


Diagram of the human heart illustrating the heart valves which may be damaged by RHD.

"It's not just about what happens in the operating theatre, but it also involves good triage, timely intervention, echocardiographic detailed assessment, outreach clinics, nursing input for family education, post-operative case audit, and more."

Finucane et al, Priorities in cardiac surgery for rheumatic heart disease, 2013.[236]

Table 30: Common valve lesions in RHD

	REGURGITATION ('leaking' or incomplete closure of the heart valve)	STENOSIS (narrowing or tightening of the heart valve)
MITRAL VALVE Valve between the left atrium and the left ventricle. The mitral valve is the most commonly affected in RHD.	MITRAL REGURGITATION (MR): the mitral valve does not close properly, causing backflow of blood from the left atrium to the left ventricle. MR is the most common manifestation of RHD, particularly in young people.	MITRAL STENOSIS (MS): mitral stenosis generally develops in more advanced RHD and is often caused by persistent or recurrent inflammation of the mitral valve.
AORTIC VALVE Valve between the left ventricle and the aorta	AORTIC REGURGITATION (AR): occurs when the aortic valve does not close properly. AR generally causes left sided heart failure. Narrowing and scarring of the aortic valve can cause obstruction to left ventricular outflow.	RHD is a rare cause of aortic stenosis.
TRICUSPID VALVE Valve between the right atrium and right ventricle	Rarely, RHD can cause isolated damage to the tricuspid valve, generally regurgitation.	
PULMONARY VALVE Valve between the right ventricle and the pulmonary artery	The pulmonary valve is very rarely damaged by RHD.	

21. MEDICAL MANAGEMENT OF RF AND RHD

THINGS TO CONSIDER

- How can primary care staff refer people with suspected RF for definitive evaluation?
- How do you ensure that people with newly diagnosed RF receive appropriate education?
- Do you have a protocol for managing RF?
- Does your programme have a management pathway for RHD?
- Does your programme have the appropriate medication and equipment to manage RF and RHD?
- How does your programme provide or refer care for people dying of RHD?

Diagnosis of RF

Diagnosis of RF is difficult worldwide, and is particularly challenging in low resource settings.[127] There is no definitive diagnostic test for RF – instead a cluster of clinical and laboratory findings relate to the probability of someone having RF. These signs, symptoms and results were codified into the Jones Criteria in 1944 to define diagnosis of RF.[581] The Jones Criteria have undergone a number of revisions since then to reflect changing needs of sensitivity and specificity. The Jones Criteria were most recently updated in 2015 to reflect the risk of RF in different populations.[13] The clinical criteria are developed by a committee convened by the American Heart Association – they are free to download online.

The updated Jones Criteria include new details about the diagnosis of RF in the era of echocardiography. In high resource settings echocardiography may allow for increased accuracy in diagnosis. In low resource settings where echocardiography is not readily available, full assessment according to the new Jones Criteria may be impossible. At a 2017 meeting of paediatric cardiologists concerns were raised that 'lack of resources make it an overwhelming task to implement the modified Jones Criteria'.[543] However, the Jones Criteria allow for a diagnostic category of 'possible' rheumatic fever category: 'In some circumstances, a given clinical presentation may not fulfil these updated Jones criteria, but the clinician may still have good reason to suspect that ARF is the diagnosis. This may occur in high-incidence settings where, for example, laboratory tests for acute phase reactants or for confirmation of recent streptococcal infection are not available, documentation of clinical features is not clear, or the history is not considered to be reliable. In such situations, clinicians should use their discretion and clinical acumen to make the diagnosis that they consider most likely and manage the patient accordingly.'[13] The guidelines allow for re-examination in 12 months with a view to making a more definitive diagnosis. The Sudan RHD guidelines use an additional category of 'probable RF' to reflect this scenario.[386]

Specialist review – by a doctor, paediatrician or cardiologist – is often needed to make a definitive diagnosis of RF. It may be useful for your programme to have a protocol for diagnosis of 'suspected RF' and 'confirmed RF', allowing primary care staff to seek specialist input, investigations and evaluation for possible cases of RF. Protocols will be needed for the referral of suspected cases for specialist investigation.

Wherever possible, specialist evaluation should occur during the symptomatic phase of RF. Some of the tests required for diagnostic confirmation (evidence of Strep A infection and markers of inflammation) can only be taken and interpreted within a short window of time after initial symptoms. Having clear pathways to complete these tests (if available in your setting) increases the likelihood of accurate diagnosis. In some places this may require admission to hospital to observe symptoms and await blood test results.[140,347,375] In other places the high burden of RF and limited access to clinical services necessitate outpatient work up of possible RF. In this case admission may be reserved for young people with symptoms of severe carditis.

Some resources have been developed to assist clinicians diagnosing RF. The Australian guidelines have been adapted to a mobile phone application which helps clinicians work through a diagnostic algorithm.[582]

A generic overview of roles and referral pathways appears in Table 31. Terms, referral criteria, resources and institutional models should be adapted to your setting.

Management of RF

The clinical management of RF should involve diagnostic confirmation, eradication of Strep A infection, management of symptoms, review for complication, education about the diagnosis and planning for follow up care.[46,583] Symptom management may include medication for joint pain or to manage abnormal movements associated with Sydenham's chorea.[583] Clinical guidelines can help to standardise this process and may be adapted to your local setting.

A small number of people with RF will have severe valve involvement which does not respond to medical therapy and may need acute surgical intervention.[236] Outcomes from acute surgery may be poor given the effect of heart inflammation (carditis).[24] In countries with access to acute surgical intervention you will need to establish criteria for referral for urgent surgical consideration.

The first episode of RF is a critical period for sharing information, education and building a foundation for long term secondary prophylaxis.

Ms Lennah Ndung'u sharing her story of living with rheumatic fever as part of the RHD Action Small Grant awarded to the Young Professionals Chronic Disease Network (Eldoret Chapter, Kenya).

Table 31: Potential roles for each level of the health system for management of RF

PRIMARY CARE*	SECONDARY HOSPITAL	TERTIARY HOSPITAL	QUATERNARY
Suspected case of RF identified. • Refer for secondary evaluation. • Register notified of suspected case. *Outpatient, primary care-based investigation of RF may be more practical in some settings.	Consider admission and specialist evaluation. • Definitive diagnosis made. • Register notified of diagnosis. • Referral to tertiary centre if evidence of heart failure.	Admission for advanced medical management. • Clinical management of heart failure. • Referral to surgical centre if required.	Admission if acute surgery required.

Table 32: Consideration following diagnosis or on discharged from RF admission (Adapted from: Australian[347] and New Zealand Guidelines[375])

TREATMENT	• Give the patient the first dose of secondary prophylaxis. • Provide a prescription for pain relief from arthralgia if still required.
EDUCATION	Broad education to include: • Explanation of RF and RHD. • Importance of secondary prophylaxis. • Symptoms that may represent a recurrence. Provide pamphlets and educations resources where available.
REFERRALS Seek formal consent when needed to share clinical details.	• Notify the case to the notifiable disease authorities in settings where RF is a notifiable condition. • Notify the RHD register coordinator of a new person to receive secondary prophylaxis. • Collect and record as may contact details as possible, including cell phone number of family, usual village and key community contacts. Provide these details to the register as able. • Contact the primary health clinic. • Arrange a dental review where possible. • Consider a referral for contraceptive services if needed for female patients.

Diagnosis of RHD

The diagnosis of RHD may be made at any stage in the causal pathway of the disease from subclinical disease detected on echocardiographic screening to advanced valve disease as outlined in Figure 2. Some people with RHD are diagnosed during a recurrence of acute RF.[584] Others are only diagnosed when they present with complications of RHD – including collapse during pregnancy or stroke. The most common presentation of RHD in endemic settings is with heart failure. Heart valve damage from RHD causes permanent changes to the pumping of blood around the heart (see Table 30 for a summary of common valve lesions in RHD). Over time, this abnormal heart function stops the heart from pumping properly causing progressive activity limitation and breathlessness. This is reflected in a study from Uganda, describing newly diagnosed RHD patients most commonly presenting with palpitation, fatigue, chest pain and breathlessness.[585]

It may be possible or necessary to make a diagnosis of RHD based on clinical history and examination. Diagnostic criteria for RHD have been described.[53] Echocardiography confirms the diagnosis of RHD and provides invaluable information about severity of disease. A detailed review by Saxena outlines echocardiography findings in the setting of symptomatic RHD for clinical staff.[150]

Management of RHD and complications

Medical management of RHD involves using medications to control symptoms, minimise disease progression and reduce complications. Education and support for people living with RHD is also an important component of management, allowing people with RHD to make informed choices about optimising their own health and engage with ongoing follow up. Surgical management may involve referral and pre-operative evaluation where surgical services exist.

Heart failure

Heart failure is the most common consequence of advanced heart valve damage from RHD. Diagnosis of heart failure may be possible from clinical examination; though ECG, chest xray, echocardiography and possibly some blood tests that can improve diagnostic accuracy.[144,586] The use of simplified echocardiography protocols to improve diagnosis and management of heart failure shows promise in low resource settings.[170] Locally adapted guidelines to manage heart failure in resource-limited settings are increasingly available.[587,588]

Management of heart failure includes the use of medications to improve heart function and medication to improve symptoms by reducing fluid overload. Access to these essential medicines is poor in low resource settings and advocacy to improve supply may be part of the role of the RHD control programme.[144] Education and support to manage heart failure (including weight measurements, dietary change, fluid restriction) are also likely to be useful but are poorly evaluated in low resource settings.[589]

In endemic settings, admissions for heart failure caused by RHD are a significant driver of health system utilisation. In Ghana, 23% of heart failure admissions to a medical ward were caused by RHD.[590] Overall, RHD is the second leading cause of heart failure admissions throughout the African continent.[591] Some of these admissions occur when people do not take, or cannot access, their regular medications – further emphasising the importance of education, access to essential medicines and universal health care.[592] Admissions for management of RHD and heart failure may be prolonged, up to 3–4 weeks in sub-Saharan Africa.[237]In low income countries the outcomes from these admissions tend to be poor – in a register study, people in Africa had a 1 year, all-cause, mortality of 33.6%.[593] Systemic barriers to accessing care, medication and education are likely to contribute to these poor outcomes in symptomatic disease.[594] Therefore, the focus on heart failure management should be early diagnosis, medical management and consideration of surgical interventions as outlined in Chapter 23.

"Importantly, when compared with other regions in this study, patients in Africa were much younger, more symptomatic, more often treated with digoxin, had little education, low rates of health insurance, and were more often from a rural area. Similar patterns were observed in India. These were the countries with highest mortality."

Dokainish et al, Global mortality variations in patients with heart failure, 2017. [593]

Atrial fibrillation

Atrial fibrillation (AF) is a common heart rhythm disturbance associated with RHD which causes the heart to beat irregularly. This abnormal heartbeat can cause symptoms, precipitate heart failure and increases the risk of stroke. In the REMEDY study of 3343 people living with RHD 21.8% also had AF.[490] Similarly, in a study of people presenting to emergency departments with AF in Africa 21.5% had a background of RHD, rising to 31.5% in India.[595]

Atrial fibrillation can sometimes be detected by clinical examination and characteristic pulse findings. Screening for AF with low cost ECG techniques may also be appropriate in some settings.[596] Devices for prolonged monitoring of heart rhythm to detect intermittent AF can also improve diagnostic accuracy.[597] The diagnosis of AF should be confirmed by electrocardiography (EKG/ECG), although access is generally limited in low resource settings. Early diagnosis of AF allows for improved clinical management and may improve outcomes.[598]

Clinical management of AF includes the use of medications to slow the heart rate, improve the heart rhythm and reduce the risk of stroke. These medications are variably available in low resource settings. Anticoagulation to reduce the risk of stroke is a particular challenge and is addressed in more detail in Chapter 22. The importance of anticoagulation in AF may be underestimated by clinicians and people living with RHD.[5] Clinical guidelines which are relevant to low resource settings are needed to improve diagnosis and management of AF.[599]

Stroke

People living with RHD have an increased risk of stroke. This may be caused by abnormal heart rhythm (AF), emboli from infection of heart valve (endocarditis) or blood clots around mechanical valves. The prevalence of RHD is likely to contribute to stroke burden in low resource settings.[600]

Stroke may be diagnosed by clinical history and examination.[601] Neuro-imaging (CT or MRI scans of the brain) can confirm the diagnosis and provide information about prognosis. In Africa, availability of neuro-imaging for stroke confirmation is variable.[602]

People with RHD are generally younger than other people who experience stroke and have an increased risk of death and disability.[603,604] The economic, social and functional impact of young people who experience stroke in association with RHD in low resource settings can be devastating. Stroke rehabilitation services can improve outcomes and function but are generally not available in developing countries.[605] Novel models of providing stroke rehabilitation, including family led rehabilitation, have not demonstrated improved outcomes. Therefore, the mainstay of stroke management in low resource settings is prevention and risk reduction.[606] The primary approach to reducing stroke risk for people living with RHD is the use of anticoagulation medication, discussed in detail in Chapter 22.

Endocarditis

Endocarditis is bacterial infection of a heart valve. Damaged heart valves are more likely to become infected which means people with RHD are at increased risk of endocarditis. The symptoms of endocarditis may be vague, leading to delays in presentation and diagnosis. Fevers and changed heart murmurs can indicate endocarditis. Echocardiography is needed to confirm the diagnosis.

Antibiotics given through the vein over many weeks are needed to treat endocarditis. Heart surgery may also be required. Prolonged intravenous antibiotics and acute heart surgery are rarely available in countries with a high burden of RHD and the outcomes from endocarditis are generally poor. For example, in New Caledonia – a Pacific Island with a high burden of RHD – 21.6% of people with endocarditis died during their hospital admission and 42.9% died during the follow up period, averaging 28.8 months.[607] Similarly, in an Australian population with access to heart surgery 20.2% of people with endocarditis died during their hospital admission.[608]

The difficulties in effective management of endocarditis requires a focus on prevention. The major source of bacteria causing heart valve infections is from the mouth. Therefore, good dental hygiene is important for reducing the risk of mouth bacteria causing endocarditis. Dental care and dental hygiene are generally poor in the places where many people live with RHD.[609] Providing education about mouth care, resources for teeth brushing and access to dental services may reduce the risk of endocarditis for people with RHD. Antibiotic prophylaxis may also be needed before dental procedures which could cause the spread of bacteria from the mouth. Details on antibiotic prophylaxis for people with heart valve disease should be covered in local clinical guidelines.

Women with RHD

Women with RHD have an increased risk of heart failure, cardiac collapse and poor fetal outcomes during pregnancy.[610] Therefore, all women of reproductive age should have the opportunity to discuss their fertility plans as part of routine health care. Many women are fearful about the impact of RHD on future pregnancies or may feel unable to ask questions. Some women living with mild RHD may be able to have a safe pregnancy with suitable medical monitoring. However, for some women with advanced heart disease, or those who have had heart valve replacement, many are advised not to become pregnant. This can have an enormous social, cultural and emotional impact and should be addressed clearly and sensitively.[611] A framework for preconception counselling for women with heart disease in Africa has been developed and should be used to inform local practices.[182]

Women with RHD who wish to delay pregnancy or cannot safely become pregnant need accurate information and access to contraception. There are major gaps in delivering essential contraception to women with advanced RHD:

- In the REMEDY registry study (conducted in 25 hospitals in 12 countries throughout Africa, India and Yemen) fewer than 4% of women of reproductive age with RHD were on contraception.[490] The majority of patients with RHD in REMEDY had moderate-to-severe valve disease and far more women had an indication for contraception than had access.
- In Malaysia, a case record audit revealed that approximately half of women with RHD were given education about avoiding pregnancy but only half of those were referred for the provision of contraception.[612]

Clinical resources to support the choice of contraception in women with heart disease have been developed.[613,614] It may be valuable to include some of this information in local clinical guidelines.

Women with RHD who do become pregnant also need information, support and planning for delivery. Many women with RHD have a poor understanding of their heart disease and implications for pregnancy.[615] In some places combined specialty clinics provide shared pregnancy and cardiac care.[184,185] In South Africa, this kind of shared care clinic has provided good outcomes for mothers and babies – even in the settings of complex heart valve disease.[184] A similar programme, RESCUE (Reproductive Services and Cardiovascular Health) is underway in Mozambique to provide integrated care for women with heart disease.[616]

Mortality and palliative care

Even with best medical therapy some people with RHD will die of their disease. In resource limited settings many people die of RHD and its complications. Their care and comfort should also be considered as part of a comprehensive RHD control programme. In low resource settings without access to surgical services the average age of death from RHD is young – in the REMEDY follow up study of 2960 people the mean age of death was 28.7 years.[5]

> "Palliative care is an approach that improves the quality of life of patients and their families facing the problems associated with life threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment and of pain and other problems physical, psychosocial and spiritual."
>
> WHO, National Cancer Control Programmes, 2004.[617]

Palliative care is the most appropriate way to manage people dying of untreatable RHD, and should focus on symptom alleviation. Severe and distressing breathlessness is a common feature of end stage heart failure. Morphine and other opiates may be used to reduce the sensation of breathlessness. Management of other symptoms at end of life may also be required.[618] Your programme should consider where people with end stage RHD should be cared for, and who will be responsible for their management. In a small number of settings, hospice or other end of life facilities may be available, though they are often focused on end of life cancer care.[619] Resources for delivery of community-based palliative care are available online.[617] Your programme may also choose to provide support for families affected by deaths from RHD.

Wherever possible, the deaths from RHD should be recorded in official mortality data or vital statistics. In places where vital statistics records are incomplete it may be possible to record deaths on the RHD register

Comorbid disease

When a person lives with two or more diseases (morbidities) these are disease are called comorbid. Management of one disease can sometime impact the management of other diseases. This is often the case in RHD, particularly as advances in medical therapy mean that people live longer with different kinds of medical conditions.

Management of RHD may be affected by a range of comorbid conditions and may need to be considered when planning service delivery models in some settings.

HIV – in the REMEDY register study, 1180 of 3343 people living with RHD had been tested for HIV infection. Of these people, 4.7% tested positive reflecting a high rate of comorbid disease.[620] This has practical implications for management – people living with both RHD and HIV in Uganda identified multiple medications and appointments as a barrier to secondary prophylaxis adherence.[621]

Similarly, medication interactions can complicate management, particularly the combination of anticoagulation with antiretroviral therapy.[622]

Ischaemic heart disease – in some countries damaging diet, smoking and lifestyle behaviours associated with ischaemic heart disease (IHD) occur in low income communities. These vulnerable communities may also be those with an ongoing burden of RHD, causing a double burden of comorbid RHD and IHD. For example, in Australia 40% of people having heart valve surgery for RHD also required a coronary artery bypass graft (CABG) for IHD. In contrast, only 21.2% of people having heart valve surgery unrelated to RHD required CABG. People with RHD were also more likely to be current smokers and have hypertension.[623] The association between RHD and IHD is less clear in lower resource settings including studies from China and India.[624-626] However, as the risk factors for IHD change in developing countries, the burden of comorbid RHD/IHD may increase. Health services may need to respond to comorbid disease by integrating IHD risk reduction into RHD care programmes, including smoking cessation messages and medical management of other risk factors.

Education and support for people living with RHD

Being diagnosed with RHD can be a frightening or overwhelming experience, particularly if the diagnosis is made late in the disease when people have symptoms or complications from RHD. However, without a strategy for consistent communication and education many people remain unsure about living with RHD and its consequences.[352,627]

Support to understand the disease, potential treatments and other health promoting activities is an important part of helping people with RHD live with their disease and contribute to managing their own care.[24,541] This information and support can be provided in different ways – general awareness raising activities are discussed in Chapter 11 on Community Education. Other specific opportunities for people who have been diagnosed with RHD are outlined in Table 33.

Table 33: Opportunities for building awareness of RHD for people living with RHD

TEACHING BY HEALTHCARE WORKERS AND WRITTEN MATERIALS	Most guidelines emphasise the responsibility of clinicians to provide education and information to people living with RHD at the time of diagnosis – including a conversation about the disease and sometimes provision of written information.[352] However, this single interaction is insufficient to communicate key messages about living with RHD over a lifetime.
TECHNOLOGY-BASED TEACHING	Different kinds of technology can be used to communicate key messages about RF and RHD. For example, in Uganda the Nurse-In-Charge of the Uganda Rheumatic Heart Disease Registry developed a group phone messaging service to communicate with people living with RHD.[271]
PEER SUPPORT GROUPS	RHD support groups have been established in range of communities, including in Kenya and Uganda.[628,629] Plans are also underway to develop an 'RHD Patient Ambassador Programme' in Namibia allowing people living with RHD to take a leadership role in developing communication messages.[630] New research is helping to quantify the benefits of support groups for people living with RHD. In Gulu, Uganda, 42 children with RHD participated in at least 3 support group activities.[566] The group met for two hours, monthly for 6 months. At the conclusion of the programme, participants demonstrated significantly improved knowledge about RHD and quality of life scores. The programme included group discussions, games, coloring in activities and viewing of echocardiography images.
EVENTS AND ACTIVITIES	Focused group events for people living with RHD provide an opportunity for sharing information about the condition to be combined with alongside peer support. One model is the 'Listen to My Heart' events run by RHD Action. Held in a variety of settings these workshops allow people living with RHD to use a stethoscope and listen to their own heartbeat, to look at Strep A bacteria under the microscope and to look at different types of prosthetic heart valves.[631] In New Zealand, a Health Youth Priority Event (HYPE) provided information for young people with RF, including details about secondary prophylaxis injections and question and answer sessions.[630] In Fiji, a wide range of events and activities are held to 'understand the purpose and benefits of secondary prophylaxis and to reduce feelings of isolation, as expressed by the patient and carer community during consultations'.[630]

22. ANTICOAGULATION

THINGS TO CONSIDER

- Are people in your programme prescribed warfarin anticoagulation?
- Are there facilities to test INR?
- Where do people have their INR tested?
- Who is responsible for adjusting the dose of their medication?
- Do you have anticoagulation guidelines?
- Are health workers trained to manage anticoagulation and its complications?

Anticoagulants are medications which make blood less likely to clot (coagulate). Anticoagulation is indicated for the management of symptomatic RHD for some patients with arrhythmia (particularly atrial fibrillation) and heart failure. People who have had mechanical heart valve replacements also depend on effective anticoagulation for survival.[22,632] Delivered effectively, anticoagulation stops clot (thrombus) formation which could cause a stroke or block a metallic heart valve.[632] In the REMEDY study (of 3343 people living with RHD in 12 African countries, India and Yemen) 40% of participants had an indication for anticoagulation.[620]

Although anticoagulation is an important element of caring for people with advanced RHD, it is often not available in the settings of greatest need. Establishing a rigorous and reliable programme of anticoagulation prior to offering surgical interventions – particularly mechanical valve replacement – is critical for developing a safe and ethical surgical programme.

A baseline assessment of anticoagulation capacity can be supported by the RHD Action Needs Assessment Tool which includes a data collection tool on anticoagulation criteria and management.

> "We collect a lot of money to get surgery for RHD patients—valve replacement costs about $5,000—but they come back a few months later with a haemorrhage, because the anticoagulant levels were not controlled, or a stroke, because they did not take the anticoagulants at all."
>
> Ali, Rebuilding the rheumatic heart disease progamme in Sudan, 2013.[371]

Vitamin K antagonists

Vitamin K antagonists (VKA) are powerful anticoagulation medications. The most commonly used VKA drug is called warfarin (and has a variety of trade names around the world). Metabolism of warfarin varies between and within individuals and is affected by genetics, diet and the use of other medications. This makes it difficult to predict how much warfarin someone will need to take to have a therapeutic effect. To account for this, most people who take warfarin require regular blood tests to measure therapeutic effect (International Normalised Ratio; INR) and adjust the dose as required. This is called 'dose adjusted to INR'.

Adjusted dose warfarin, titrated to INR is difficult to manage, even in relatively high resource settings. Management requires a very high level of numeracy and health literacy for both health workers and people living with RHD. Management of warfarin anticoagulation is often calculated as the time-in-therapeutic range (TTR). There are a number of methods of calculating TTR, the simplest of which is the number of INR tests in range divided by the total number of tests.[633]

Delivering safe, effective and reliable anticoagulation is a worldwide challenge:

- In an Indigenous Australian population, one third of RHD patients on adjusted dosed warfarin had inadequate warfarin monitoring.[634]
- In Kenya, in a cohort of patients with RHD and atrial fibrillation, only 52% of INR tests were in the therapeutic range over 12 months of follow up.[635]
- In Nairobi, in a hospital based cohort of people with RHD, INR was therapeutic on only 40% of clinic visits over a 12 month period.[636]
- In a register based study of people living with RHD in Namibia, only 39% of people who needed anticoagulation were taking Warfarin and 73% had no INR recordings in the preceding 6 months.[637]

Inadequate INR monitoring and variability of TTR is associated with very poor clinical outcomes, including high risk of stroke and catastrophic valve thrombosis.

INR monitoring

The major issue with warfarin is medication adherence and monitoring of INR.[632,638] The target INR depends on the indication for warfarin, for example, in valve replacement, arrhythmia, etc. The dose of warfarin needs to be changed (titrated) in accordance with the INR. There are a number of models for monitoring INR and titrating warfarin dose outlined in Tables 34 and 35.

Various protocols and algorithms are available to guide adjustment of warfarin dose based on INR. These may be included in national resources as in South Africa and Rwanda.[127,641] There is reasonable evidence to show that use of this kind of algorithm to adjust warfarin dose improves time in therapeutic range.[642]

Table 34: Models of INR testing

LABORATORY INR TESTING	In the laboratory model, patients have a venous blood sample taken, either in the clinic or at the laboratory. The blood test is processed in the laboratory, INR measured, results are provided to health professionals and then instructions are provided to the patient about the dose of warfarin to take. Results take time to process and communicate, sometimes arriving days after the initial bloods test. In some countries, variable quality control of results and high laboratory costs are barriers to laboratory-based INR monitoring.[639]
POINT OF CARE INR TESTING	Point of care testing (POCT) is a new approach, allowing patients or health workers to measure INR on a small machine and receive a rapid result. POCT has been adopted in a number of low resources settings and has made it possible to decentralise INR monitoring.[127,640] Machines require occasional testing/calibration and ongoing supply of reagent cartridges which can be expensive. Recommended technical specifications of POCT machines are available online.

Table 35: Models of warfarin dose adjustment

ANTICOAGULATION IN PRIMARY CARE	In places where primary care is delivered by medical staff, anticoagulation monitoring is commonly arranged through primary care.[643] This allows primary care doctors to provide integrated comprehensive care for a range of medical conditions, and maintain frequent contact with people needing close INR or clinical monitoring.[643]
SPECIALIST ANTICOAGULATION CLINICS	In some low resource settings anticoagulation is considered a specialty service and delivered in dedicated tertiary clinics with staff skilled in dose titration and experienced in responding to INR fluctuations.[635,644] This specialised focus may allow for safe and effective anticoagulation.
PHARMACIST LED ANTICOAGULATION	Anticoagulation education, dosing and monitoring may be arranged through pharmacy services.[164] This kind of task shifting may facilitate more frequent patient engagement and satisfaction.[645] Depending on the skill and resource mix in your setting it may be possible to develop an integrated anticoagulation programme with a chemist or pharmacy.
PATIENT LED ANTICOAGULATION	Some highly health literate patients in well-resourced settings have their own point of care INR machines and adjust their own warfarin dose within pre-specified limits.[646] Growing access to point of care INR machines means this approach is increasingly feasible in developing settings – including a nurse led, patient performed INR service for people living with RHD in Haiti.[647]

Other medications

Aspirin

Aspirin may be sufficient for stroke prevention in some patients with heart failure or low risk atrial fibrillation. Your RHD management guidelines should include advice on when aspirin is an appropriate option for anticoagulation. The Partners in Health guidelines from Rwanda are a good example of this kind of triage.[127]

Heparin

Heparin is a short acting anticoagulation medication which is administered as an injection (either as an intravenous infusion or as a low-molecular weight form via a subcutaneous injection). Heparin may be used to provide anticoagulation for pregnant women because it does not have the same risks to fetal development as VKA medications.[648,649] Low molecular weight heparin was added to the WHO Essential Medicines List in 2015.[650] However, in low resource settings low molecular weight heparin is largely unavailable or prohibitively expensive.[611]

New oral anticoagulants (NOACs)

New medications for anticoagulation, which do not require blood test monitoring, have been developed and are increasingly widely used for a variety of indications. However, these NOAC medications are not currently recommended for anticoagulation in the context of RHD.[651] A clinical trial, INVICTUS, began in late 2016 to explore the safety and efficacy of NOACs in people with RHD and atrial fibrillation in low resource settings. Results from the trial are expected to be available in 2020.[652] The NOACs should not be used for patients with mechanical heart valves.[653]

Education when initiating anticoagulation

When anticoagulation is started, patients should be provided with enough information to take the medication safely and communicate important information to other clinical staff. High quality anticoagulation is a lifesaving intervention for people with mechanical valve replacement. However, preliminary results from the multicentre REMEDY study of people living with RHD suggest that only 35% of people on warfarin know their target INR.[654] Understanding of INR and warfarin doses is further complicated by low numeracy in some settings.[655]

In one study from a Pacific Island, 1 in 8 people with mechanical heart valve replacements for RHD had stopped taking anticoagulation.[656] People who reported they did not understand the need for ongoing medication were more likely to have stopped taking warfarin.[656] Therefore, education at initiation of warfarin anticoagulation and during future clinical encounters is required. A systematic review of anticoagulation education components (mainly in developed countries) identified the following domains for discussion:[657]

- Basics of anticoagulation.
- Risk-benefit and the indication for anticoagulation.
- Adherence (including the strength of different tablets, their different colours and what to do if doses are missed).
- Accessing health care (when to seek medical attention).
- Diet (including foods which can impact the metabolism of anticoagulants).
- Laboratory monitoring (including target INR).
- Medication interactions.
- Self-care (including management of bleeding, planning of pregnancy, sporting participation).
- Access to INR testing services.

Anticoagulation medication which some people with RHD need to take daily to prevent stroke

A large number of handouts and other resources are available online to help communicate information about anticoagulation. However, these may need to be adapted to provide critical information in settings of low literacy, numeracy or low health system resources. The Cardiological Society of India has developed good practical guidelines on anticoagulation education following mechanical valve surgery.[658]

Anticoagulation education may also be task shared with pharmacists. In Sudan, an education programme of pharmacy information and written materials for people taking warfarin for heart valve disease improved knowledge of the medication and resulted in improved adherence.[659] In China, a pilot programme of inpatient education led by pharmacists while people recovered from heart valve surgery showed some promise as a communication model.[660]

Supporting adherence

Many of the barriers to medication adherence addressed in Chapter 18 for secondary prophylaxis are also applicable to anticoagulation. Inadequate education about the need for therapy, cost, health beliefs, inconvenience and travelling distance, all contribute to poor anticoagulation adherence.[661] Strategies to address some of these barriers may include providing people with an INR record card. Such records can provide a useful way to communicate the target INR, date of next test and required dose.[662] This approach is widely used in high resource settings, Rwanda and in the Pacific Islands.[321,663]

"At discharge, patients are provided with a booklet used to record their INR values, medication lists, and other comprehensive information. Patients are asked to bring this booklet at each subsequent visit to ensure optimal care and promote continuity of care from inpatient to outpatient management."

Patton-Bolman, Developing a sustainable model for cardiovascular care in Rwanda, 2015.[663]

23. TRIAGE OF INTERVENTION CANDIDATES AND PRE-OPERATIVE EVALUATION

THINGS TO CONSIDER

- How does your manage the list of people waiting for surgery?
- Does your programme have a relationship with a regular surgical or interventional service?
- How do you communicate to providers about potential surgical candidates?
- How do you begin to prepare patients for the experience of surgery and secure informed consent?
- How do you investigate co-morbidities and ensure that people are medically optimised before surgery?

There are a number of models for accessing surgical services for people living with RHD, potentially including a mix of international evacuation, visiting surgical teams and local surgery. These are outlined in more detail in Chapter 25. Even in settings with limited surgical access it may be helpful to identify people who should be considered for surgery (surgical candidates). Developing a surgical waiting list can provide an signal of unmet surgical need and facilitate better screening and preparation of surgical candidates should care become available. Systems are also needed to ensure that people are medically, mentally and emotionally prepared for intervention – factors that are important for ensuring the best possible outcomes.

This chapter provides an overview of pre-operative issues for individuals, and for the health system. The next chapter (Chapter 24) addresses post-operative considerations for individuals, surgical teams and health services. You should consider both chapters before interventional services (Chapter 25) are delivered.

Mitral valve disease (regurgitation and/or stenosis) is the most common pathology of RHD. Although other valves and heart structures may be damaged, mitral valve procedures are the most frequent interventions for RHD. Refer to Table 30 for an overview of common valve lesions in RHD.

Table 36: Overview of surgical procedures in RHD

MITRAL VALVE REPAIR	Mitral valve repair is an open heart surgical procedure. Surgeons repair the shape and function of damaged valve leaflets allowing for more normal blood flow. Repair offers the best possible outcomes for children and adults with RHD and is associated with lower short and long term mortality.[664] However, repair surgery is less durable and often requires reoperation. Surgical techniques for repair are technically more difficult than valve replacement.[236]
VALVE REPLACEMENT	Heart valve replacement is an open heart surgical procedure. Surgeons remove the damaged heart valve and replace it with a mechanical prosthetic (metallic valve) or bioprosthetic valve (tissue valve). Bioprosthetic valve replacements cause fewer blood clot complications than metal valves but are more likely to wear out and require replacement. Mechanical valve replacement is associated with high risk of embolism and haemorrhagic complications but usually last for life. Children who have a heart valve replaced may need repeat surgery as the heart grows.
BALLOON VALVOTOMY (VALVULOSPLASTY, COMMISSUROTOMY)	Balloon valvotomy is used in some settings for the treatment of mitral stenosis. This closed surgical approach (percutaneous) is used to open a narrowed mitral valve by gently inflating a balloon inside the valve. The procedure may need to be repeated some years later. Clinical outcomes have been positive in the African setting,[494] and in the Indigenous Australian context.[71] The closed approach reduces costs and complications compared with open surgical repair, providing a safe and effective option for low resource settings.[494] Importantly, women with mitral stenosis who are pregnant may be able to have the procedure ahead of delivery. However, a cardiac catheterisation laboratory is required to perform the procedure and few facilities exist in the areas of greatest need.
HEART TRANSPLANT	In a small number of settings heart transplant is available for people with advanced heart valve disease which cannot be improved with valve-specific surgery.[665]

Table 37: Pre-operative investigations

ECHOCARDIOGRAPHY	Echocardiography data provides critical information regarding valve lesions, cardiac chamber size, left ventricular function and pulmonary artery pressure; and serial data will assist with determining the timing of surgery.[450] Information about pre-operative left ventricular dysfunction also provides information for risk stratification, improving the information available to inform consent processes.
DENTAL OPTIMISATION	Dental optimisation prior to heart valve surgery reduces the risk of subsequent bacterial endocarditis and is a standard part of pre-operative preparation in most settings.[672] In Australia, inadequate dental preparation was one of the reasons planned rheumatic valve surgery was postponed and the patient returned home.[673]
NUTRITION	People living with RHD are at risk of under-nutrition and growth stunting.[674] A person with a good nutritional status pre-surgery will have improved post-surgery outcomes compared with a person who is undernourished.[22,675]
PREGNANCY STATUS	Female surgical candidates being evacuated or travelling for surgery should have their pregnancy status confirmed before departure. Pregnancy is not necessarily an absolute contraindication to intervention but should be considered prior to travel.
INFECTIOUS DISEASE STATUS	Evaluation for potential infectious diseases will vary by setting but may include testing for tuberculosis, HIV, hepatitis B, hepatitis C and malaria.[127,672,674]
ROUTINE PRE-OPERATIVE BLOODS	Full blood count, liver function tests, creatinine, glucose, electrolytes are routinely checked in most settings before surgery.[127,672,676]
BLOOD GROUPING	Blood must be available for transfusion during and after heart surgery. Identifying the blood group of the patient and cross matching of blood may need to be organised in conjunction with relatives in some settings.[672]
EVALUATION FOR ISCHAEMIC HEART DISEASE	It is standard practice in some settings for older or high risk patients to undergo coronary angiography prior to valve surgery. This allows for identification of co-morbid ischaemic heart disease which may change management plans.[677]

Pre-operative issues for the health system

Triage and waiting list management

Any consideration of cardiac surgical capacity – locally or international – should begin with pre-operative planning. For the health system, this means a local consensus decision about who should be referred for surgical evaluation, timing of referrals and the process of evaluation. Ideally, everyone with symptomatic heart valve disease should be evaluated to consider whether surgery would relieve symptoms or improve outcomes.[586]

In practice, considerations of who can be offered surgery will usually include patient factors and health systems factors, including:[127]

- Capacity of individual patients to benefit from surgery. This may include consideration of the optimal timing of surgery according to age and clinical status.
- Experience and ability of the surgical team.
- Post-operative ward capacity.
- Training needs of local surgeons.
- Cost of surgery.
- Access to required follow up, including anticoagulation and secondary prophylaxis.[236,658,666]

A relationship between locally-based health care staff (from hospitals or the RHD control programme), and surgical teams, is required to build trust, improve handover and monitor outcomes. Where possible, each case should be discussed between clinicians – including adherence with anticoagulation and BPG post-operatively, plans for pregnancies, degree of functional impairment and follow up arrangements.[667] There are a number of detailed clinical guidelines to inform these discussions. WHO offers some clinical and echocardiographic indications for surgical referral.[22] In Australia, all symptomatic patients with clinical congestive heart failure are considered for intervention.[667] In Rwanda, cardiac surgical section is coordinated nationally by the cardiac surgery programme director and colleagues.[127]

The use of a priority-based care planning system, as outlined in Chapter 19, will assist with the triaging of candidates for rheumatic cardiac surgery, as those with moderate to severe levels of valvular lesions will have been monitored and reviewed more frequently and more data will exist regarding the patient. Political interference in triaging referrals is a challenge in some countries.[321] Transparent criteria for referral may help address this issue.

Estimating surgical volume

Planning for interventional care should include an approximate estimate of the number of people who may benefit from pre-operative assessment. This can help inform triage systems, waiting list management, quantify unmet demand and support advocacy for improved access to interventional services.

A model to estimate disease progression has been developed using real data from 617 Aboriginal and Torres Strait Islander people living with RHD in Australia. In that population, 50% of young people with severe RHD at diagnosis required surgery within 2 years of that diagnosis.[668] As this data is drawn from a high-income setting with reasonable access to advanced surgical services the volume of services delivered is a reasonable indication of clinical need for surgery. Most people in developing countries present with advanced heart valve disease.[5,6] Therefore it is reasonable to assume that at least half of people newly diagnosed with RHD would be candidates for surgical assessment were resources available. In highly endemic settings the proportion of newly diagnosed people living with RHD who should be considered for surgery is likely to be even higher. In the VALVAFRIC study of 3441 people diagnosed with RHD in eight countries, 83% required surgery. Only 2.2% were able to receive the heart surgery they needed.[6]

Even when some surgical services are available the demand for surgical services is large, generating long waiting lists. For example, in Vietnam more than 5000 adult people living with RHD are awaiting surgery.[669] In Rwanda, 2000 people with RHD and congenital heart disease were on the surgical waiting list in 2015.[670] Ensuring that waiting lists are managed, particularly when there are multiple surgical service providers locally, visiting and internationally, can be difficult. In Haiti, a centralised register of people awaiting heart surgery had helped manage referrals and improve overall access to care.[647]

Clinical preparation

People living with RHD and being triaged for intervention may well have other health conditions or comorbidities. A pre-operative period with structured and systematic medical evaluation is good practice and will allow a balanced risk assessment to be undertaken. Accurate clinical information and clear communication with the patient supports the informed consent process (outlined in more detail later in this chapter).[671]

Considerations for pre-operative optimisation in your setting may include:

Your programme will need to discuss with your surgical team(s) – local, international or visiting – the role of each of these pre-operative investigations and decide:

- Who will decide which tests are indicated for each patient?
- Who is responsible for arranging each investigation and following up results?
- How will results be recorded and communicated to the surgical team?

Ideally, high quality pre-operative evaluation will occur in local settings, rather than having patients travel to tertiary centres and then be identified as unsuitable surgical candidates.[236] Pre-operative preparation should be started early but is often conducted a few weeks prior to visiting surgical teams arriving.[678] This is one role that local teams may be able to lead as part of capacity building towards in independent surgical services discussed in Chapter 25.Optimising pre-operative assessment has been an important element of reducing post-operative mortality for visiting surgical teams performing paediatric congenital heart disease surgery.[679]

Education and informed consent for surgical candidates

Informed consent is the process of medical staff providing patients with information about potential treatment options and associated risks and benefits before making a decision about treatment.[680] There is no agreed international definition of exactly what is considered informed consent, though ethical, legal and administrative requirements are often specified.[681] The approach and attitude of surgeons to informed consent also vary, particularly in developing countries.[682] Literacy, traditional and religious beliefs in developing countries and within sub-groups in developing nations also influence the nature of informed consent.[683]

Ultimately, the principles of providing information to people living with RHD about their disease, treatment, risks and expectations, underpin good clinical practice. RHD control programmes which are facilitating access to cardiac surgery should seek out local guidelines for informed consent standards. For example, detailed guidelines have been developed in South Africa and India.[684] In settings without an established standard for informed consent RHD control programmes may need to facilitate development of informed consent processes for cardiac surgery.

Surgery for the management of rheumatic heart valves is often frightening for patients – particularly when the proposed intervention is to be delivered in a distant setting or country.[504] The process of obtaining informed consent takes time and ideally begins long before the date of surgery. Discussions should begin early, allowing individuals and families to make a meaningful decision about the pathway forward. This should include information about the surgery itself, the risks and benefits of the procedure, recovery time and the requirements for ongoing follow up. In some places this may need to incorporate a discussion of the costs of ongoing care.

For the person to be informed and able to provide valid consent they need to be provided with information that is understandable to them, therefore the health practitioner should take into account the health literacy of the person, consider various ways to communicate the messages and utilise a variety of materials. For example, in Sudan, an evolving paediatric cardiac surgery programme included diagrams of the heart for patients' counselling and Parents' Information Pamphlets designed using simple, local (Arabic) language, and which included diagnosis and treatment plans.[685] In Rwanda, pre-operative education modules have been developed to provide information to surgical candidates and families.[663]

Pre-operative planning for people living with RHD

The role of the surgical candidate preparing for surgery varies in different countries. Considerations which may need to be discussed include:

Anticipated costs of surgery: In most low-resource settings families or sponsors need to pay for surgical services.[676] The time needed to collect these funds can be a delay to surgery.[663,672] Providing families with estimated costs is an important element of surgical planning in some settings.

Arranging for blood donations: In some settings family replacement donors are required to access blood banks. If this is the case, it should be discussed and planned ahead of the surgical procedure.[686]

A/Professor Liesl Zühlke and a member of the South Africa Community Advisory Group for RHD examine a prosthetic heart valve at a Listen to My Heart event.

24. POST INTERVENTION REVIEW, FOLLOW UP AND AUDIT

THINGS TO CONSIDER

- How do other local/visiting surgical services follow up patients in your setting?
- How and when will responsibility for care transition back to usual services?
- How will post-operative patients be followed up for clinical, and outcome monitoring?

'Surgical outcomes' generally refers to mortality after surgery, major complications of surgery and the need for repeat operations. Outcome may also include quality of life and return to preoperative level of function. Surgical outcomes for RHD vary worldwide, often with poorer outcomes in low resource settings. Although this may reflect late presentation of advanced heart disease it is increased by difficulties following up patients, maintaining anticoagulation and identifying post-operative complications early.[667]

Establishing a robust structure for post-operative follow up can improve surgical outcomes. Planning for the post-operative period should be considered prior to delivering intervention services. This helps to ensure that people receiving the intervention get the most benefit, that limited funding is used appropriately and that heart surgery is delivered in the safest possible environment.

Follow up is important to optimise outcomes of individual patients and to understand outcomes from surgical services.

> "Late postoperative care has proved to be one of the biggest challenges because many of the adult patients are taking warfarin for anticoagulation after receiving mechanical heart valves. Financial constraints around clinic and hospital visits and admission have prevented some patients from receiving appropriate care in a timely fashion, again, most critically, in the vulnerable first few weeks after surgery."
>
> Yankah et al, 'Cardiac Surgery Capacity in Sub-Saharan Africa', 2014.[687]

1. Post-operative planning for the health system

Diagnosis and treatment guidelines for Strep A infection and secondary prophylaxis guidelines help to standardise care. Similarly, a structured pathway of care around the time of a heart operation is also needed. The aim is to improve the quality and safety of care provided which meets the needs of individuals, families and your local capacity for service delivery. An overview of post-operative logistic considerations is provided in Box 25.

Establishing a schedule of post-operative visits may minimise confusion and ensure that follow up expectations are consistent between clinical staff, patients and families. The model should be developed in conjunction with the primary health sector to support transitions between care. Follow up requirements generally involve wound care, medication adjustment, regular echocardiography and blood tests to monitor for side effects of cardiac medications.[688]

It may be possible to decentralise late post-operative follow up to primary or secondary care (See Table 38 for guidance on planning for 'step-down' care after cardiac surgery). This allows people to return closer to home during recovery. However, staff in secondary and primary care facilities should be able to identify, manage and refer conditions associated with a range of post-operative complications for this to be safe. This should include recurrent symptoms, fever, evidence of heart failure, new murmurs, thromboembolic episodes, signs and symptoms suggesting endocarditis. Specific post-operative training may be required. Written information about surgical procedures and post-operative plans should be kept by people who have surgery if referral systems between different levels of the health system are poor. It may also be necessary to include post-operative follow up for people who have had heart surgery internationally in routine follow up systems.

Table 38: Planning for 'step-down' care after cardiac surgery

PRIMARY CARE	SECONDARY HOSPITAL	TERTIARY OR QUATERNARY HOSPITAL
• Monitoring of complications • Repeat prescriptions/regular medications • Ongoing education and support for patients and families	• Potential for 'step down' or convalescent care • Management of complications • INR monitoring	Cardiac intervention performed • Discharge education • Anticoagulation initiated if required • Follow up appointments scheduled

BOX 25:
Post-operative logistic considerations

- How long will patients be expected to stay near a tertiary setting post-operatively?
- Is there funding or accommodation support once discharged from hospital?
- Will the patient be able to receive the level of care required locally post-operatively?
- What process will be in place to ensure the providers of care at the patient's local health facility are informed and provided with relevant clinical information?
- Is there a protocol in place at the local facility to ensure the primary health workers understand the routine care required?
- Will the tertiary health service providers provide ongoing care in the form of routine reviews?
- Who will local health care providers contact if they have concerns?
- Will peripheral health centres or chemists have a supply of post-operative medications prescribed on discharge?
- Are there telephone support services or a hotline for people living remotely to call with concerns?

2. Post-operative planning for individuals

In addition to planning for heart surgery, people with RHD should be supported to plan for life after heart surgery. This generally involves a hospital stay, a period of recovery/rehabilitation and ongoing medical follow up. Post-operative planning should occur far in advance of surgical procedures, be addressed when informed consent is secured, and be reinforced during the hospital stay and at every post-operative visit. Caregivers and families should be involved in these discussions as much as possible.

Hospital admission

People in hospital for cardiac surgery may have a prolonged admission. Parents and caregivers are often required to stay and care for hospitalised children, making it difficult for parents to work or care for other children. Some people will travel overseas for heart surgery, necessitating additional logistic considerations. Providing information to families about the estimated duration of the hospital stay and potential timing of surgery may be helpful for planning.

The inpatient period provides an opportunity for education, particularly around specific issues at the time of hospital discharge. This may include pain management, wound care, necessary blood tests and information about potential complications. Children's HeartLink have developed a parent education/discharge instructions (PEDI) resource for the delivering of pre- and post-operative education related to children's heart surgery. This is available in a variety of languages and low literacy settings.[689] Use of PEDI-associated resources appears to have improved nursing confidence in discharge education in a low resource facility in India.[690] The benefits of providing education on discharge care may be significant – for example, family education on wound care may reduce surgical-site infection.[691]

> "We emphasise patient/parental education before discharge. This effort is directed by a clinical pharmacist led team. We focus attention on the need for regular monitoring and control of anticoagulation, food and drug interactions with warfarin, and prophylaxis for both endocarditis and rheumatic fever. We generally require patients to report for follow up every 4 weeks."
>
> Edwin et al, The development of cardiac surgery in West Africa – the case of Ghana, 2011.[692]

Recovery and rehabilitation

A period of structured outpatient cardiac rehabilitation following heart valve surgery is an important part of recovery in high resource settings.[693] It is far less common in low resource settings but may have an important role in providing information, social support and return to full function. Cardiac rehabilitation may be available for young people who are transferred for surgery in high resource settings. For example, children from Zimbabwe having surgery in Italy participate in a rehabilitation programme followed by some months of convalescence with host families.[694] A structured format for post-operative advice and follow up should be considered in planning for providing surgical services.

Ongoing medical care

People and families should be given enough information to plan for ongoing medical care after surgery long before the operation actually occurs. This may include anticoagulation (discussed in Chapter 22), other medications and regular cardiac follow up. This care may be an out-of-pocket cost for people with RHD and their families so information about anticipated costs of services should be addressed.

The need for ongoing follow up is not always clear to people in the post-operative period. People who have had severe breathlessness and exercise limitation with severe RHD may experience significant symptomatic improvement after surgery. In some cases this is misinterpreted as a 'cure' and patients sometimes mistakenly stop all medications.[695,696] Communication with patients and families is essential to explain that surgery is not a definitive solution for RHD.

> "Those parents are seeing their child healthy and they think that they don't need any medication anymore."
>
> Tchoumi et al, Surgical management of cardiac valvular lesions in a tertiary Sub-Saharan centre, 2012.[695]

Quality of life for people who have had heart valve replacements has been described in a number of developed countries. In general, quality of life is improved after surgery and these improvements are sustained over a number of years.[697] The impact of heart surgery on quality of life for people in developing settings is starting to be explored. For example, in Rwanda, women and people living in rural villages had lower post-operative quality of life scores than other people who had heart surgery.[698] Understanding quality of life of people who have had heart surgery provides important information about education and health system needs to support outcomes of people living with RHD. As articulated by one of the surgical pioneers of mitral valve repair techniques, 'It's not enough to save patient lives, we must also take into consideration the quality of life given to the patient and the socio-economic impact of our surgical actions.'[699]

3. Post-operative planning for the surgical team

> "Although all aspire to provide high-quality care, outcomes evaluation should be an integral part of every program, especially when services include invasive procedures with the capability to harm as well as help."
>
> McQueen et al, The provision of surgical care by international organisations in developing countries: a preliminary report, 2009.[700]

All surgical services (local, visiting and international) should be able to measure their post-operative outcomes. This allows services to be able to give risk-stratified information for informed consent, ensure practice is consistent with local/international standards and to inform ongoing improvement.

Increasingly robust standards for post-operative outcome recording have been developed for congenital heart disease surgery for children in low resource settings. For example, the International Quality Improvement Collaboration for Congenital Heart Surgery in Developing World Countries (IQIC) reports surgical outcomes from 58 sites in 24 countries and over 62,000 surgeries.[701] The IQIC has made it possible to focus on areas of surgical care which can be optimised to improve outcomes, including the reduction of surgical site infections, team care and nursing engagement.[701] Review of surgical outcomes data from a single site database can also inform practice changes.[679] Although focused on congenital heart disease, this approach is also relevant to surgery for RHD in similar settings by similar teams.

Establishing a framework for ongoing audit – including standardised data collection forms and recall schedules – should be embedded into surgical planning. In the absence of standardised international database for RHD surgery, a local outcomes database should be developed prior to provision of cardiac surgery. Consultation with surgical programmes in the region to develop this database may support data harmonisation and make it possible to benchmark local outcomes.

25. PROVISION OF INTERVENTIONAL SERVICES

THINGS TO CONSIDER

- Are there any existing cardiac surgery services accessible to your setting?
- Is there political and clinical leadership to increase these services or establish independent capacity?
- Are there visiting teams or nearby surgical programmes who can provide advice?

Capacity for cardiac surgery in settings with a high burden of RHD is very limited. A variety of approaches for delivery surgical services have evolved to address this unmet need. These models are summarised in Table 39 and addressed in more detail in the following sections. This chapter is intended to provide a framework for discussion of different models of service delivery and is not a comprehensive overview of technical issues. Countries beginning new cardiac surgery programmes, engagement with visiting teams and international training of local staff should consult widely with technical experts before adopting new strategies.

These models of care may co-exist within the same country. For example, in a 10-year study in Cameroon, 63% of children who had heart surgery travelled overseas. The remaining 36% had their operation through the Chantal Biya Foundation in Yaounde – some performed by local surgeons and others by visiting surgical teams.[702] Similar mixed-model arrangements have existed in Jamaica and in Rwanda.[663,703] In these locations, it is particularly important to have strong pre-operative and post-operative planning so that care for people having surgery through different pathways can be efficiently coordinated.

> "Although cardiac surgery is an important player in the building of cardiovascular disease control, it may prove insufficient as a stand-alone strategy. The number of initiatives taken by NGOs has been growing over the past 20 years. Political will to improve healthcare systems globally is needed to render these programmes sustainable."
>
> Mirabel, Cardiac surgery in low-income settings: 10 years of experience from two countries, 2017.[704]

Table 39: Models of cardiac surgery in endemic RHD settings

INTERNATIONAL SURGICAL TRANSFERS	VISITING SURGICAL TEAMS	INDEPENDENT NATIONAL OR REGIONAL CENTRES OF EXCELLENCE
(+/- international training of staff from low resource settings)		
Some countries have no local capacity for cardiac surgery. In these cases, people living with RHD may have the opportunity to have surgery internationally through government-sponsored or philanthropic programmes.	In some resource-limited settings, visiting surgical teams provide intermittent cardiac surgical services.	Some larger low and middle-income countries have been able to work with visiting surgical teams to develop independent cardiac surgical units.

International surgical transfers

In settings without any access to cardiac surgery it may be possible for people with RHD to have surgery internationally.

International surgical transfers can happen formally through the health system. This is the case in a number of smaller countries, particularly in the Pacific Islands.[453] Historically, most of the surgery for RHD in the Pacific Islands was performed in New Zealand or Australia, creating informal regional 'centres of excellence'. For example, in New Zealand up to 50% of operations in the single paediatric cardiothoracic unit are on patients from overseas, particularly the Pacific Islands.[236] There are similar arrangements for government supported (and partially privately funded) transfers for children needing heart surgery in Namibia to travel to South Africa.[237]

International surgery may also be arranged through charitable programmes. Some of these are structured bilateral programmes, such as the patient transfers from Timor-Leste to Australia.[705] This programme is based on pre-operative planning, focused case selection and capacity building to improve delivery of RHD care in Timor-Leste.[705] Other models involve people living with RHD being transferred to a variety of different international hospitals for surgery. For example, children from Cameroon have travelled to France, Italy, Switzerland, Belgium and South Africa for cardiac surgery.[702] People with RHD from Rwanda have travelled to both India and Sudan.[663]

Programmes arranging international transfer for surgery have inherent limitations – the volume of surgery performed is almost always insufficient with little opportunity for scale up, the process does not facilitate knowledge transfer and continuity of care is limited.[694,706] The impact of international travel for a major medical procedure in an unfamiliar environment with vast differences in culture and language on people living with RHD is also unclear.

Visiting surgical teams

A 2013 survey identified 80 non-government organisations (NGOs) providing paediatric cardiac care services in 92 low and middle income countries.[707] In addition, other NGOs support adult cardiac services. Therefore, these programmes are a major contributor to global cardiac surgery services in developing countries. Visiting teams are the only option for cardiac surgery in a number of RHD endemic countries. These services generally cost much less than procedures performed internationally in high resource settings.[708] However, humanitarian missions have potential risks and benefits – for visiting teams, patients and local staff.

There are considerable benefits from international cardiac surgery missions, including opportunities for knowledge transfer with local staff, raising awareness of RHD and delivery of much needed surgery which would not otherwise be available.[707,709] However, visiting surgical services share the major limitations of international transfer for surgery – the impossibility of visits being frequent enough to really meet population need. As described in the Ugandan setting, 'relying on visiting surgeons and sponsors is not sustainable because the number of patients they can operate on during a brief visit is limited and falls short of demand. Also, patients' conditions worsen day by day and many die before the next available visiting surgeon.'[710]

Supporting the development of sustainable, independent cardiac services is one of the most important elements of surgical visits.[703,706] However the benefits of knowledge transfer and health system strengthening only occur when they are deliberately planned elements of each trip. Simply travelling overseas and performing surgery does not, in itself, support health system strengthening and improvements in care. Benefits can only be meaningfully accrued when visiting services align with local priorities, foster relationships, teaching and goals setting.[698] Visiting teams with an exclusive focus on clinical volume are a missed opportunity for knowledge sharing and capacity building.[711] Some large countries have had decades of visits from cardiac surgical teams without ever growing local capacity for surgery.[237]

Various stepwise models for development of cardiac surgical centres in low resource settings have been proposed, though most focus on the process of establishing partnership programmes from the perspective of the visiting team.[708,712-714] Programmes initiated by lower resource settings tend to have a slightly different focus, outlined in the next section of this chapter.

Visiting surgical teams face a wide array of challenges and have significant potential for harm.[712] Challenges include staffing, equipment, hospital infrastructure, communication and language barriers, poor engagement with surgical candidates and financial limitations.[708]

Harms associated with visiting cardiac surgery teams can be considerable. For example, in 2008 a visiting surgical team from New Zealand travelled to Samoa to provide heart surgery. Fourteen operations were performed (13 for RHD) and the visiting team departed four days post-operatively. Two patients died within 30 days and six were re-admitted following discharge with pericardial effusions.[453] Similarly, procedural changes prompted by visiting teams can have unintended consequences, for example by driving greater surgical volumes than local settings can accommodate or disrupting access to necessary surgical facilities.[715] Some of these risks can be mitigated through close relationships with local teams, careful planning and thoughtful attention to health systems issues. For example, a checklist of ethical considerations for international surgical missions has been proposed and may provide a foundation for reflection.[716]

Although many visiting teams provide valuable, capacity enhancing services which support a transition to independent services, other programmes do not. As observed from South Africa 'Most of Africa relies on flying paying patients, or donor-funded patients, to centres off the continent, or hosting short-term visits of skilled personnel'. There is a marked lack of coordination in the latter and some NGOs have not learnt the lessons of sustainability.'[237] A global strategy for planning, coordinating and evaluating visiting cardiac programmes to low resource settings is urgently needed, with an emphasis on local capacity building and community-accountable models.[707]

Development and expansion of local surgical services

Ideally, cardiac surgery should be delivered in settings which are geographically and culturally close to countries with a high burden of RHD. This is reflected in the World Heart Federation Position Statement on RF and RHD which identifies the need to 'increase the capacity for cardiac surgery in countries where RHD is endemic' as a priority.[227]

It is difficult to measure exactly how many centres provide cardiac surgery in settings with endemic RHD. A 2012 survey of facilities in Africa identified 78 sites performing regular open-heart surgery in the continent. A total of 10,725 heart surgeries were performed across these sites in 2012, of which nearly a quarter were for RHD.[687]

A number of these programmes have published accounts of developing capacity for RHD surgery, successes and ongoing challenges. These generous descriptions make it possible to identify common themes and provide a foundation for discussion about developing cardiothoracic surgical capacity in endemic settings. This overview in Table 40 is not exhaustive but illustrates the broad scope of surgical experience in low and middle-income countries. Other programmes are well underway but have published fewer descriptions of programmatic issues, for example in Algeria,[717] Morocco,[718] Tanzania,[719] Cambodia,[704] and elsewhere.

The technical requirements of establishing sustainable cardiac services are outside the scope of TIPs and best provided by peer support from existing programmes. The details, logistics and choices involved in providing cardiac surgery for RHD in low resource settings require ongoing political support, financing and programmatic support from experienced centres.

Table 40: Illustrative overview of cardiac surgery services for RHD in low and middle income settings

GHANA[692]	The National Cardiothoracic Centre of Ghana has been operational since the 1990s with financial support from the governments of Ghana and Germany. Initial funding allowed staff from Germany to stay in Ghana for 2–10 years. The Centre is now one of the few independent cardiac units in West Africa providing services by resident, locally trained, staff. Between 2002–2011, 1775 heart operations were performed, 21% for RHD.[720] Out of pocket costs are high for people requiring surgery and financial sustainability is an ongoing challenge. The centre is accredited by the West African College of Surgeons as a training location and more than 20 surgeons from neighbouring countries have been trained.
CAMEROON[721]	The Cardiac Centre of St Elizabeth Catholic General Hospital in Shisong, Cameroon, was inaugurated in 2009. People in Shisong who needed cardiac surgery had previously needed to travel to Italy. Between 2009 and 2011, 23 visits have been arranged to provide cardiac surgery in the new facility. Between visits, resident doctors offer medical management, catherisations and pacemaker implantations. Surgical services attract a significant out of pocket cost and families of many children with RHD can not afford surgery.[722] There are also outstanding gaps in education for people living with RHD and provision of secondary prophylaxis.[722]
JAMAICA[703]	A reinvigorated programme of cardiac services began in Jamaica in 1994, supported by 41 visits from seven different visiting surgical teams through to 2011. Most operations are now performed by local surgeons, a small number which are for RHD. A memorandum of understanding including the Ministry of Health was signed in 2010 to develop a self-sustaining paediatric cardiac centre.
RWANDA[52]	A novel collaborative partnership has been possible in Rwanda – the Rwanda Cardiovascular Care Consortium – bringing together four different visiting surgical teams, the Rwanda Ministry of Health, Rwanda Heart Foundation and four Rwanda cardiologists to achieve common goals.[52] Training placements for local cardiac surgeons have been addressed. The programme has an increasingly comprehensive focus on RHD control including expanding surgical capacity, strengthening register-based secondary prophylaxis and improving primary prevention.[698]
UGANDA[145]	A paediatric cardiothoracic programme was restarted at the Uganda Heart Institute in 2007 following a period political upheaval. A number of visiting teams supported training and service delivery for lower risk congenital heart disease surgery. Local surgeons began performing independent congenital surgery in 2010. For RHD the focus has been to develop appropriate support services, including secondary prophylaxis and optimised medical management. The plan is to scale up surgery for RHD. The Government of Uganda has supported the development of paediatric cardiac surgery along with a large number of strategic partners. There is an ongoing unmet need for heart surgery for RHD for adults and children.[723]
SUDAN[724]	A number of hospitals offer cardiac surgery in Sudan, some supported by government funding. Visiting teams and philanthropic contributions support service delivery – predominantly for RHD valve disease.[724] Since 2004, specialist paediatric cardiology services have been available at the Sudan Heart Centre and embedded in a programme of team building, increased echocardiography capacity, database building and development of clinical protocols.[685]
EGYPT[301]	The Aswan Heart Centre opened in 2009 with the support of an international surgical team and local philanthropy. Adult and paediatric heart surgeries are offered. A strong focus on skills transfer has allowed the percent of procedures performed by local surgeons to increase from 2% in 2009 to 74% in 2014. Full time staff employment has been credited as creating a more sustainable model.
MOZAMBIQUE[725]	The first open-heart surgery was performed in Mozambique in 2001 by a visiting team from Europe. The initiation of open-heart surgery followed collaborative efforts by a Mozambican NGO and four humanitarian surgical providers from Europe in partnership with the Ministry of Health in Mozambique. International training of staff from Mozambique and epidemiologic research on the burden of RHD have been major elements of the programme. The team from Mozambique now provide surgical some surgical services between trips of visiting surgical teams.[704] Supply of essential medical equipment and out-of-pocket costs for people needing heart surgery is an ongoing challenge.

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