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RHEUMATIC HEART DISEASE
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Tools for implementing rheumatic heart disease control programmes

TIPS Handbook

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Introduction

Each year, nearly half a million 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, 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 a simple 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 affliction in most parts of the world. The majority of sore throats are short viral infections which resolve without complication. However, a substantial minority of sore throats are caused by a bacterial infection. The most common cause of bacterial sore throats is group A streptococci (GAS). In susceptible young people GAS infections of the throat can cause an abnormal immune reaction, known as rheumatic fever (RF).[1,2] This abnormal immune response causes inflammation of the heart (carditis) and, with repeated GAS infections, scarring of the heart valves. Damage to the heart valves indicates rheumatic heart disease (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.

Nearly half a million people worldwide suffer an episode of RF each year and at least 15 million people live with subsequent valve damage of RHD.[3,4] Robust epidemiologic data for RF and RHD is insufficient; the true burden of disease is likely to be several times higher than current estimates.[4,5] Approximately half a million people die of RHD annually around the world.[3] Overwhelmingly these deaths are premature; on average, people dying from RHD are aged under 40.[6,7]

The majority of people with RHD live in developing countries.[3] Others live in high resource countries in Indigenous communities and other vulnerable populations. The socioeconomic distribution of RHD reflects its roots in poverty, overcrowding, 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.[8,9] Symptomatic RHD simultaneously reduces employability and increases health care costs. In endemic settings people living with RHD often bear the economic cost of accessing health services, medication and sometimes, prohibitively expensive heart surgery. 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 marital harm.[10,11] 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). 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.

RHD control programmes have been implemented around the world for more than fifty years. 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 and preventing the development of RHD. Delivery of these services often requires health system interventions including health worker training, government engagement, and disease notification systems. RHD control programmes may also incorporate medical management of symptomatic RHD, facilitate access to cardiac surgery, conducting research to understand the burden of disease, primordial prevention to tackle underlying risk factors. TIPs collates the implementation experiences of 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.[12] The relevance of each component will be determined by local needs, priorities and experience.

What are rheumatic fever and rheumatic heart disease?

Up to 30% of sore throats in children and young people are caused by a bacteria called **group A streptococci (GAS)**. Without antibiotic treatment some of these children will develop rheumatic fever (RF) a few weeks after their sore throat.

RF causes joint pains, fever, skin changes and sometimes abnormal movements. In most cases the heart also becomes inflamed during RF. However, when other symptoms of RF resolve, changes to the heart valves persist. Repeated episodes of GAS infection and RF cause progressive heart valve damage. This persistent valve scarring is called rheumatic heart disease (RHD).

The risk of RF following untreated GAS pharyngitis is between 0.3 - 3%.[13-15] For individuals with a history of previous RF the risk rises to 50%.[16, 17] The most important determinant of disease progression appears to be the number of times RF recurs in an individual.[18]

The classical pathway of individual progression from GAS infection to RF and RHD is illustrated in Figure 1.

This diagram is a simple way to understand the disease. Advances in echocardiography have revealed that the reality is probably a little more complex. A diagram of disease progression at a population level appears in Figure 2.[4]

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.[18] These mechanisms are relatively poorly understood and are not addressed in this handbook.

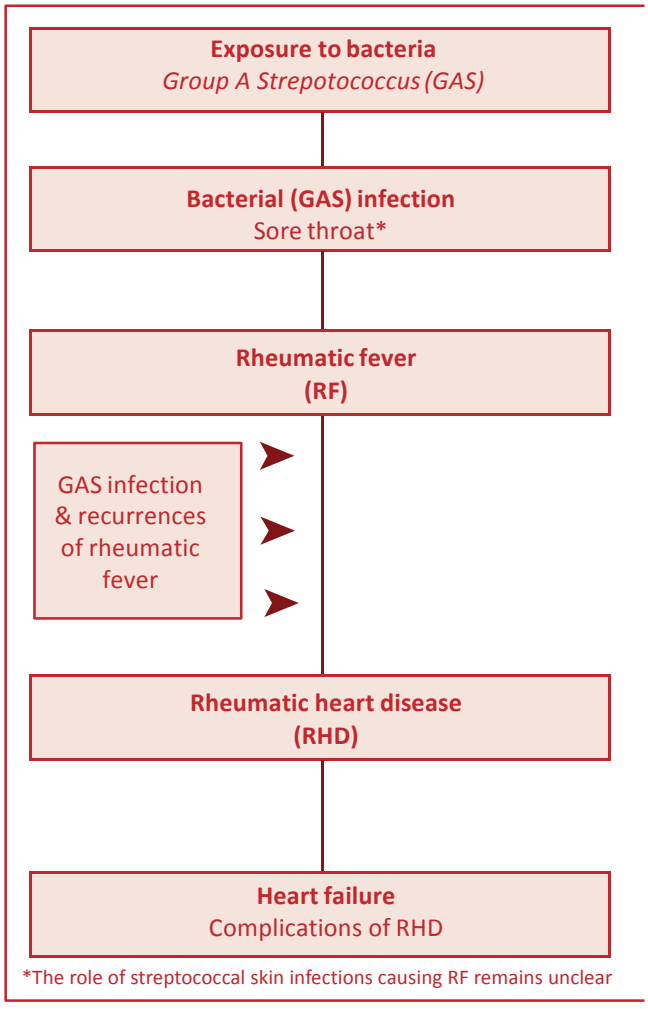

Figure 1: Causal pathway of RHD

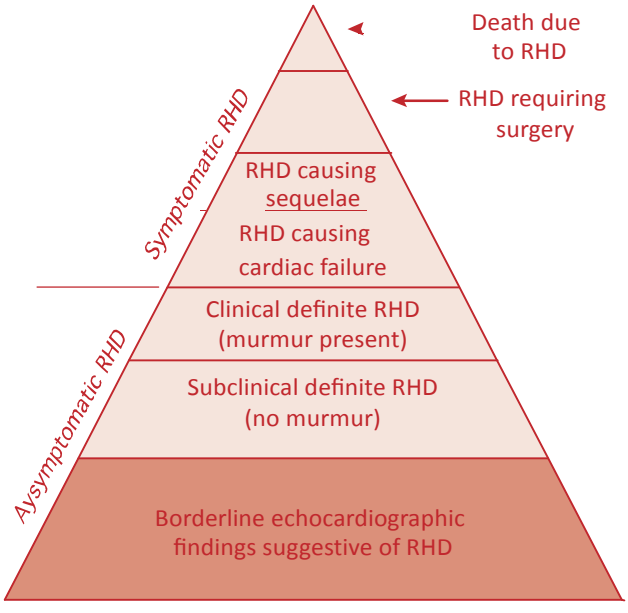

Figure 2: Population model of RHD progression[4]

What is a comprehensive RHD control programme?

There are many opportunities to intervene on the pathway from GAS 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 for many years.[19, 20] In reality, most programmes are more than a register – they include efforts to treat sore throats, educate communities, arrange antibiotic supplies and treat the complications of advanced RHD. These programmes are called 'comprehensive' because they include primary, secondary and tertiary components. The importance of this kind of multimodal approach to RHD is increasingly recognised.[5, 21]

A comprehensive approach is exemplified by the A.S.A.P (Awareness Raising, Surveillance, Advocacy and Prevention) Model developed by the Pan African Society of Cardiology **(PASCAR)**.[22, 23] The A.S.A.P model incorporates four key elements: education, primary prevention, secondary prevention and disease surveillance.[23, 24] These components offer a clearly articulated policy overview of the domains required for disease control.

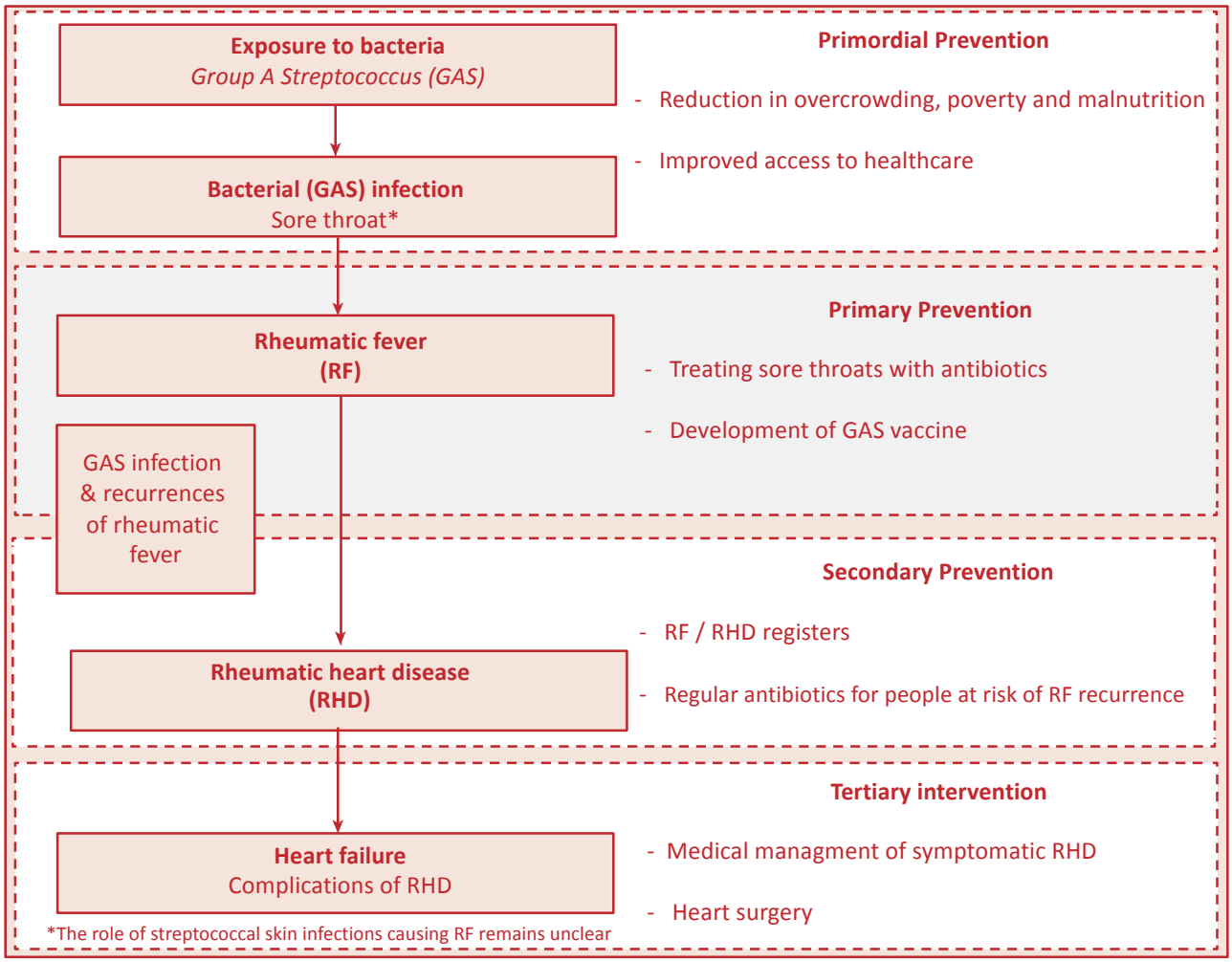

Figure 3: Opportunities for intervention in RF and RHD

Decisions and tasks required at a programme level are necessarily more detailed. In 2013 the recommended components of comprehensive RHD control programmes were collated and structured into a conceptual framework.[12] This implementation framework provides a structure for the following TIPs chapters.

An approach to describing, designing, implementing, and evaluating comprehensive RF/RHD control programmes is outlined in figure 4. The components are arranged in approximate order of priority, working from left to right, bottom to top, in each row.[12] This conceptual framework emphasises the need to tackle less complex components (antibiotic supply) before more complex interventions (echo screening and cardiac surgery). Suggested priorities for new programmes are: collection of burden of disease data, fostering government engagement, community education, development of an RF/RHD register and medical management of existing cases of RHD.

The framework is not designed to be prescriptive, your programme certainly doesn't need to tackle everything once. The details of designing and delivering RHD care will be unique in each setting. Local needs - community consultation, existing infrastructure, political and economic feasibility of programmes and human resources- are the most important considerations. 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.

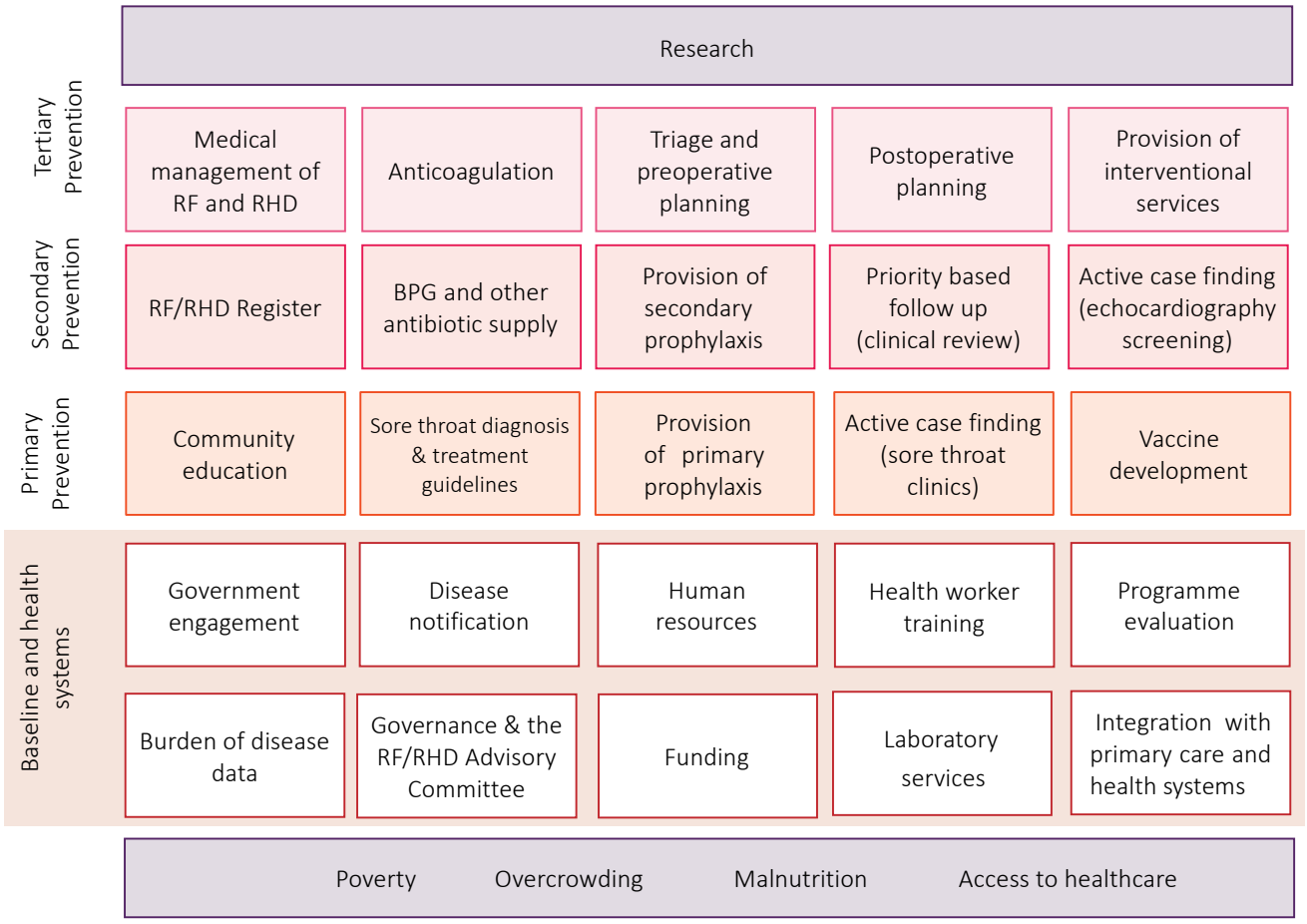

Figure 4: A conceptual framework for comprehensive RHD control programmes. Components are arranged in approximate order of priority, working from left to right, bottom to top, in each row.

How to use this handbook?

The text is lengthy 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 in different situtations. 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.

Subheadings are marked throughout TIPs:

Things to consider

Conceptual question based summary at the start of each chapter.

★ Opportunities for research

Outlines areas where your programme may be able to contribute to the global knowledge base about control of RF and RHD. There is also a 'Research' chapter at the end of the book addressing overarching research needs.

★ Opportunities for integration

Highlights scope for your programme to work with other disease communities, programmes or departments to improve care delivery. Chapter 5 is dedicated to integration in more detail.

★ Opportunities for primordial prevention

Addresses some opportunities about preventing the development of RF.

Who should use this handbook?

TIPs is written primarily for people implementing RHD 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 part of a group of people beginning to plan a control programme. You may want to evaluate an existing programme or participate in a humanitarian surgical mission.

TIPs is designed to be 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 involved in control of RF and RHD on page 11. A short version, 'Quick TIPs' has also been produced for easy reference. Other terms are in the **glossary** (marked in the text in purple), explained in boxes in the text or hyperlinked in the online version. If there are things you think should be clarified or better explained in future editions we'd love to hear from you; contact details are in the inside cover.

Methods and limitations of TIPs

TIPs collates 60 years of programmatic experience delivering different components of comprehensive RHD control programmes.

The core references were identified through a systematic literature review of EMBASE, BIOSIS and PubMed searches of English and French articles from 1952 – 2012. 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 review of institutional archives including the World Heart Federation and WHO. 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. Participants in key informant interviews were also asked to recommend other sources of unpublished grey literature.

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. Many questions critical to the management and control of RF and RHD are poorly understood. Much of the evidence comes from historical studies in relatively high income countries from the 1950s – 1970s. It is unclear whether these experiences can be directly extrapolated to currently endemic regions.[3]

- 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.

- Literature review was limited to English and a small number of French language resources. Experience from other 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 specialised disciplines in their own right. We have summarised key issues in these domains and provided additional references for further information.

It can be done!

Although control of RHD of can appear overwhelming, the achievements of landmark programmes demonstrate that significant progress is possible. Cases 1- 3, page 9 illustrate some of the achievements in RHD control made possible by comprehensive control programmes.

Case 1: The Pinar del Rio Cuban Experience

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:

"The project included primary and secondary prevention of RF/RHD, training of personnel, health education, and dissemination of information, community involvement and epidemiological surveillance. Permanent local and provincial RF/RHD registers were established at all hospitals, polio clinics and family physicians in the province. Educational activities and training workshops were organised at provincial, local and health facility level. Thousands of pamphlets and hundreds of posters were distributed, and special programmes were broadcast on the public media to advertise the project." [25]

By 1996 the incidence of RF had fallen from 18.6/100,000 to 2.5/100,000 and recurrence rate fell dramatically. This reduced burden of disease persisted until at least 2002, even when the control programme

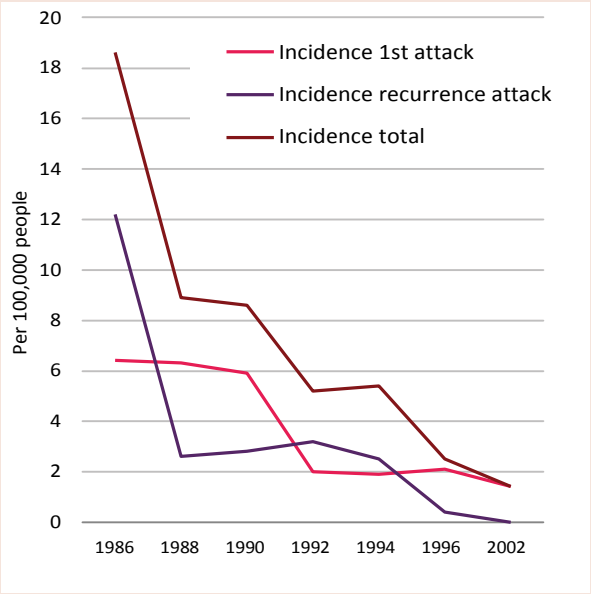

Figure 5: Incidence of RF per 100,000 people aged 2- 25 years, Pinar del Rio, 1986- 2002.[25]

Case 2: 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 with free access to health care and medication.[26]

The programme had four key principles:

1. Development of a register
2. Health worker and community education
3. Research
4. Treatment of skin infections

A full time paediatrician dedicated to RF was employed on each island. By 1992 the incidence of RF had declined by 78% in Martinique and 74% in Guadeloupe. The cost to the health systems of RF reduced by 86%.[27]

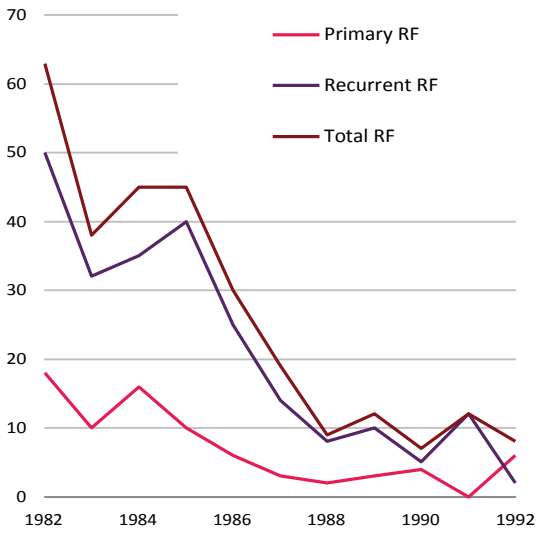

Figure 6: Number of cases of RF in Martinique 1982-1992.[27]

Case 3: The Northern Territory, Australia Experience

In 1997 the Northern Territory (NT) region of Australia developed a register based programme for control of RF and RHD.[28] RF is a notifiable condition in the region and active surveillance is maintained through intensive health professional education and dedicated programme staff. All cases of RF, recurrences and RHD are entered into a computer based register. There are over 54 remote primary health centres into addition to regional and urban primary health facilities. Each facility operates a local register with a patient recall component providing data to the NT central RHD register. In 2013 there were approximately 2500 people on the central register, the overwhelming majority Indigenous Australians (Aboriginal and Torres Strait Islander Peoples). People on the RHD register are managed according to comprehensive clinical guidelines grounded in regular injections of benzathine penicillin G.[29] Public health nurses employed by the NT RHD Control Programme travel the NT and provide support to remote health centres in the development and delivery of services. Nurses provide training and education to remote health staff, patients and their families. This service delivery is integrated with other public health programmes, such as the trachoma programme.[30] A programme review in 2013 provided evidence of programme success: the **recurrence rate** as fallen by 9% per year since the programme began in 1997.[31]

Health system structure

Levels of the health care service

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.[32] 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 below – you should define and adapt your own local terms when developing local guidelines and protocols for **referral** between health services.

Primary	Health clinics Health centres Family doctors
Secondary	Local hospitals Visiting specialists
Tertiary	Referral hospitals National hospitals
Quaternary	Specialised national or regional units Visiting services International services

Introduction to vertical, horizontal and diagonal approaches to health care

One of the ongoing debates in health care - particularly in low and middle income countries (LMIC)- is how to structure funding streams, care delivery and governance. Debate is often centered on '**vertical**' programmes (for specific diseases like HIV/AIDS, malaria and tuberculosis) versus '**horizontal**' programmes (usually health systems strengthening interventions, like improved primary care and/or laboratory services).

In reality, a combination of horizontal and vertical programmes is usually needed; sometimes called a 'diagonal' approach.[33] '**Diagonal**' programmes strengthen primary health care and infrastructure alongside disease specific activities.

Control of RF/RHD is well suited to a diagonal approach.[34,35] Horizontal health systems components may include improved access to primary care, treatment of sore throats, access to laboratory services, referral pathways and primordial prevention. Vertical components may include a register and dedicated staff to deliver secondary prophylaxis injections.

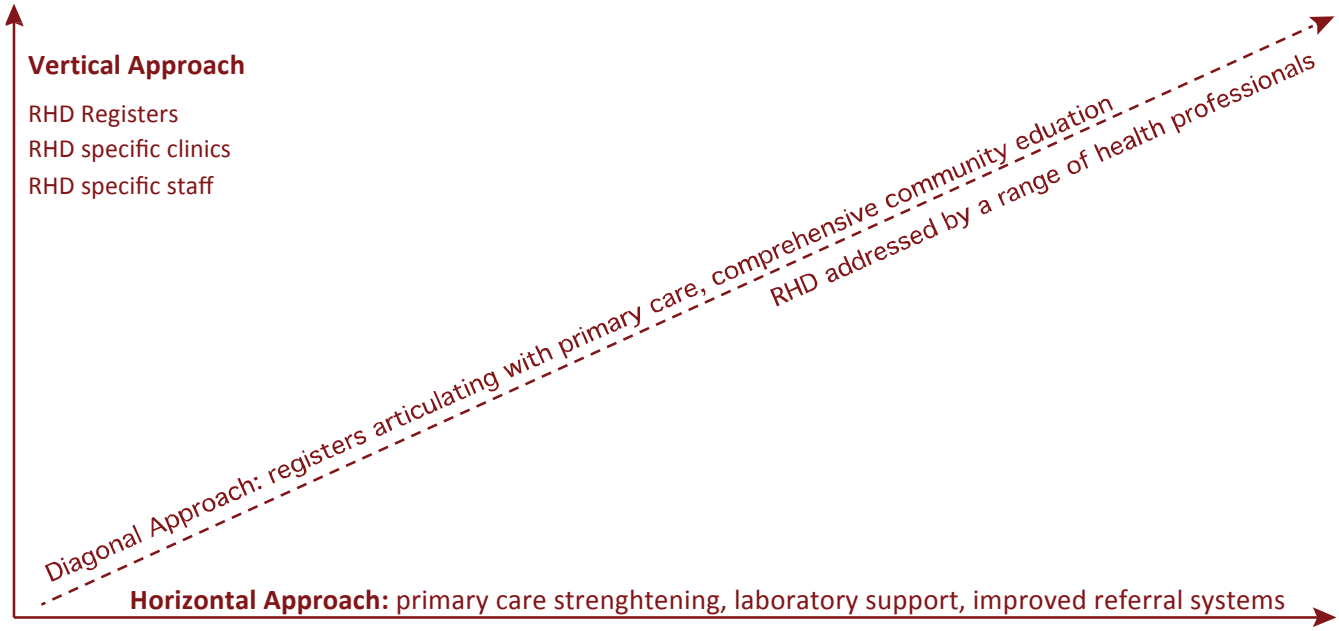

Figure 7: Horizontal approach: primary care strengthening, laboratory support, improved referral systems

Table 1: Merits of vertical and horizontal programmes[33]

	Advantages	Disadvantages
Vertical	• Targeted, allows rapid results and 'easy wins' • Outcomes easily measured/quantified • Health professionals can be trained to provide specialist services	• Fragmented experience of care. Patients may need to visit many different clinics for different needs. • 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

Clinical background

This section provides a plain text summary of medical conditions relevant to RF and RHD. It is provided to assist people without extensive clinical training to become familiar with medical issues in RF and RHD control.

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 (the mitral valve in 90% of cases) cause chamber pressures to rise, and the heart to fail as a pump. Without a well-functioning heart, fluid builds up in the lungs and body, causing symptoms like breathlessness, swelling and fatigue. These symptoms tend to become worse over time without treatment.

Stroke

A stroke (also known as a **cerebrovascular accident, CVA**) 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 attributable to underlying RHD.[3]

Infective endocarditis

Infective endocarditis (IE) is a bacterial infection on the valves of the heart. Valves that are already scarred or damaged by RHD are more likely to have IE than undamaged valves. Worldwide, approximately 60% of people with IE have underlying RHD.[3] People with IE have fevers and the heart may be unable to pump blood effectively. It can be difficult to diagnose IE and - even when IE can be diagnosed - antibiotic treatment may be ineffective. Minimising the risk of IE is an important part of managing RHD. The bacteria that cause IE tend to come from the mouth, so good dental hygiene is an important way to minimise risk. Giving prophylactic antibiotics before dental work and some other procedures is standard in some countries.

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 cause palpitations. AF also significantly increases the risk of stroke. In endemic settings RHD is a major cause of AF.[36]

Maternal morbidity and mortality

Women with RHD are at risk of significant illness or death during pregnancy and labour. 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 causing heart failure. The symptoms of heart failure may be confused with symptoms of late pregnancy and go untreated, causing cardiovascular collapse and death. Women who have received heart valve surgery and mechanical heart valves are at risk of serious bleeding from anticoagulation medication. These medications can also affect the developing baby.

Case Study 1 | World Health Organization Global Programme

Dr Porfirio Nordet, Former Medical Officer, Cardiovascular Disease Programme, World Health Organization

The World Health Organization (WHO) has been engaged in RF/RHD control and prevention efforts since the 1950's. The most substantial of these activities was the WHO Global Programme for the prevention of rheumatic fever and rheumatic heart disease, implemented from 1986-2002 in 16 pilot countries in high endemic regions throughout the world. Dr Porfirio Nordet, former advisor to WHO on RHD, shared his experiences as a facilitator of the Global Programme:

Components of the Programme

WHO provided protocols and educational materials to participating countries in addition to motivational support and modest financial backing. In exchange, countries were responsible for regularly reporting data to WHO. The programme was rolled out in phases. Phase 1 involved a single pilot site within each country. During phase 2, project expansion into the surrounding communities took place. In phase 3, 5-10 years post-implementation, programmes scaled-up to the national level. Progress reports were submitted to WHO on a semi-annual basis. The Global Programme focused on secondary prophylaxis but also attempted to implement robust primary prevention in 7 target countries. Secondary prophylaxis required the creation of a central registry and active case finding which was completed by direct survey of school students and families for symptoms as well as a review of hospital records. An unstable supply of BPG, inadequate staff and weak reporting were all associated with lower rates of adherence to secondary prophylaxis.

Public, patient and provider education was a significant component of the programme. Print media, including booklets, pamphlets and posters, were preferred forms of messaging because of their easy reproducibility and distribution. At a quick glance, they can refresh providers' medical knowledge and improve their clinical practice. Patients can be reminded of follow-up recommendations or of the natural history of their disease. Awareness campaigns also utelised the mass media through radio and television. Healthy lifestyle education and hygiene promotion were taught to young people in schools. A direct correlation was seen between the amount of education provided and the number of patients registered and compliant with secondary prophylaxis.

Challenges and Solutions

Follow-up was difficult for patients living far away. Recommendations were made to decentralise follow-up care to local health clinics. In addition, swabbing and culture were discouraged because of delays in returning test results and because patients often did not revisit the clinic to receive a definitive diagnosis. Since RF and RHD are not very common conditions for primary care physicians to manage, knowledge of diagnostic criteria and disease management decreases with time. Concise and accesible resources, like posters and pamphlets, were considered essential for both providers and patients to most efficiently manage GAS pharyngitis and RF/RHD.

A major problem was programme sustainability post-2002, after the Global Programme's funding had been disbursed. Country reporting to the WHO slowed, probably in association with cessation of a (small) financial incentive. Structural changes at the WHO and a declining sense of international camaraderie contributed to reduced engagement after 2002. Changes in leadership and approach at the national level further reduced capacity. Sustainability was closely tied to a lasting commitment of the Ministry of Health which, in turn, requires constant advocacy by local champions. From a programme's inception, champions should be sought out through Ministries of Health by identifying well-respected experts at local institutions. Champions then assemble a network of physicians and coordinate their activities with the Ministry of Health.

Dr Nordet reflects that constant evaluation, requiring a central registry and periodic reports and presentations to the Ministry of Health or international organisations should be part of any programme to promote accountability.

Ideally, protocols supplied to countries would be locally-relevant, taking into consideration the realities on the ground. To do this, WHO staff would necessarily visit countries to assess their infrastructural strengths and limitations before drafting guidelines.

Health systems & 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 be supported by a strong foundation of baseline and health systems considerations.

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 evaluate 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 for 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. At every opportunity RHD control programmes should strive for clinical and health system integration. Conscious and considered integration supports sustainability, improves care delivery and makes it less likely that RF/RHD services will be prematurely dissolved. Integration may occur clinically, in partnership with perianal care, dental services and non-communicable disease services. Integration at a health systems level may be needed to strengthen primary health care systems, include RHD in NCD action plans and foster disease notification systems.

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 population. RHD exemplifies the ideal integrated, diagonal, health care delivery in low resources settings. Well designed and delivered programmes have tremendous potential to be beacons of best practice for other disease communities.

Things to consider

Who does your RHD control programme provide care for?

What sources of denominator data are available?

What is the age distribution of the 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?

Have you documented the processes for developing your burden of disease estimate, to allow updating in the future?

1. Burden of disease data

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.[22, 38-40] Epidemiologic data is particularly important as it allows your programme to:

- evaluate whether RF/RHD is a public health priority
- provide baseline data to identify targets and monitor the impact of any intervention
- motivate governments and funding organisations to engage with your project
- know how sensitive and specific clinical tests, tools and guidelines will be in your setting

The importance of denominators

Knowing how many people your programme delivers care for provides an important denominator for 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

Understanding the total number of people you care for is also important for monitoring trends over time. If your population changes- through growth, immigration or re-zoning - 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.[40]

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

Beginning a new RHD control program may require burden of disease estimates and simultaneous development of a register (see chapter 16). Active case finding may be required to collate all sources of information: injection books, admission records, cardiology referrals and other sources of clinical information. Development of a register and estimates of burden of disease may occur concurrently.

$$\frac{1\ (\text{numerator})}{1000\ (\text{denominator})}$$

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.[40]

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.

Box 2: Beginning a new programme

"One key challenge in addressing RHD is the lack of reliable data capturing the true burden of disease, i.e. precise geographical, age and gender distribution of health systems measurements such as where and when care is being sought, by whom and at what cost."

Dr Agnes Binagwaho
Rwanda Minister of Health 2013.[42]

Sources of burden of disease data

Multiple sources of information provide signals 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. It is important that your methods for estimating incidence, prevalence and mortality are clearly documented. Without a well described approach, changes in rates may be incorrectly attributed to programme success or failure rather than changes in estimated methodology.

Hospital or health records

Most 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 will tend to underestimate disease frequency because only people who present to health services and are admitted will be recorded – potentially missing people who are unable to access health care, or those with symptoms that are too mild to seek medical aid.[19] Alternatively, tertiary or specialist hospitals often accept patients from a larger geographic area than local health services. This may cause an overestimate of the burden of disease. It is helpful to know what proportion of the hospital patients come from your target population/denominator. Understanding the frequency of RF and RHD in hospital/tertiary settings is also valuable for estimating costs of caring for a population with a high burden of RHD (See Chapter 3).

In some places, injections delivered by a health centre or hospital are recorded in an injection book. It may be possible to identify people already receiving secondary prophylaxis antibiotic injections from these books or other records of care delivery.

Death records

Details about the numbers and causes of death (sometimes called vital statistics or mortality records) are collected in almost all countries. This information can provide valuable signals about the burden of RHD. Where record keeping systems are weak, deaths may not be recorded, and significant under-reporting is common.[42] In these cases, mortality data may be useful in setting a firm 'lowest possible' burden of deaths threshold. In some places, autopsy data may provide information about the burden of RHD.[43]

Extrapolating from similar countries

Even without local data it may be possible to estimate the burden of disease from similar areas or countries. Settings with similar economic development and ethnicity are likely to provide the best guidance.

Historic estimates

Before the widespread use of echocardiography, **cardiac auscultation** was commonly used to screen schoolchildren for heart murmurs and RHD.[44] Although auscultation is now known to significantly underestimate the true burden of RHD, historical studies may provide some information about disease prevalence.[46]

Note: 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 20, Chapter 20.

Disease notification

In some places rheumatic fever is a notifiable condition, providing valuable information about the rate of disease over a period of time (see box 1, burden of disease terms). However, under reporting is common, particularly when systems to report cases are weak. RF and RHD are largely a clinical diagnosis which can make notifications susceptible to change, as a result of education or outreach activities. For example, increased awareness of diagnosis 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.

Echocardiography screening

Echocardiographic screening of school-aged children 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. The role of echocardiography in clinical management of disease is still under investigation.[46] The risks and benefits of echo screening are outlined in Chapter 20.

Global burden of disease data

Attempts have been made to estimate the burden of RHD at a global level.[3, 4, 48] Although this may be imprecise, comparing local estimates with global figures can provide a valuable check for plausibility.[3]

★ Opportunities for research

Medical, nursing and other students can be important contributors to small research projects. Review of hospital records to review burden of disease data can be a worthwhile project for students and local programmes.

★ Opportunities for integration

Vital statistics registers are essential elements of a health system. Improved mortality data allows for improved services for a wide range of conditions. In countries with weak mortality reporting infrastructure, RHD control programmes may be valuable advocates for improving data collection. Interoperability between systems is an important consideration to ensure communication between multiple data sources. Resources and information are available from WHO.[43]

Vulnerable populations

The burden of RF and RHD varies between and within populations. RHD is most common in vulnerable groups including Indigenous communities and socially and economically disadvantaged people.[37, 48]

Globally, RF and RHD are more common in Indigenous communities than non-Indigenous communities.[50] The association has been demonstrated in Canada,[51, 52] the United States,[53, 54] Brazil,[55] South Africa,[56] New Caledonia,[57] Fiji,[58] Australia[31] and New Zealand.[59] Disparity may be very pronounced. In Australia RF/RHD affects Indigenous communities almost exclusively: 97.6% of first episode RF between 1997 and 2010 occurred in Indigenous Australians.[30] Indigenous Australians are 122 times more likely to live with RHD than non-Indigenous Australians.[27] In New Zealand, 91% of people with RF are Māori or Pacific peoples.[58] Migrants and refugees may also live with RHD in high income settings where the disease is otherwise controlled.[18, 60]

This variability in distribution 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. Similarly, reporting the burden of disease with a whole population denominator may mask a significant burden of disease within subpopulations. You may 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.
- The size of vulnerable populations may need to be estimated separately with an independent denominator which may need to be drawn from alternative sources.
- 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.

Age

RF and RHD have a relatively predictable age distribution worldwide, illustrated in Figure 7. RF typically occurs between 5 and 20 years, with a peak incidence of first episode RF at 11-12 years. Symptomatic RHD can begin in childhood and prevalence increases with age.[3]

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.[40] The all-age prevalence of RHD is expected to be 5-7 times higher than the prevalence in 5-14 year olds.[3] Developing countries with a high burden of RF also have very young populations which should be taken into account when reporting on the burden of disease, 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 (usually 5 or 10 years) to reflect the variation in risk with age.[41]

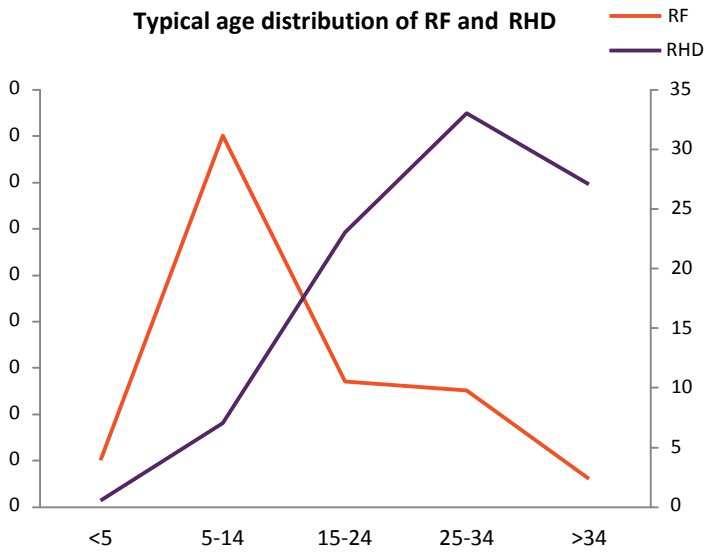

Figure 8: Typical age distribution of RF and RHD, Northern Territory of Australia[3]

things to consider

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/terms of reference?

What are their primary goals for the programme?

Ministry of Health
Ministry of Education
Ministry of Housing

- Primary care/general practice/family medicine
- Nurses/midwives and local health workers
- Medical Specialists:
  - Cardiologists
  - Paediatrics
  - Internal medicine
  - Cardiac surgeons or representatives of surgical programmes
- Dentists
- Epidemiologists
- Microbiologists
- Laboratory services
- Echocardiographers
- People living with RF / RHD
- Community leaders
- Faith based groups
- Family groups
- Schools and teachers
- World Health Organization
- Donors and funders
- National/international heart networks and societies
- Private sector partners
- International technical advisors

2. Governance & the RHD Advisory Committee

"The programme advisory committee consisted of a part-time provincial cardiologist, supported by representatives of related departments and bodies 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"

Dr Nordet, Cuba, 2008.[26]

Designing and developing an RHD control programme requires input from many different stakeholders.[21, 23, 61] The goals, strategy and planning for the programme are usually overseen by a committee of these stakeholders. Decisions are also needed throughout the programme; it is helpful to have a group of people already assembled who can provide advice and oversight.[62] An RF/RHD Advisory Committee also provides a pool of individuals who can advocate for prioritising the disease, even when comprehensive programmes are not yet established. For example, the advocacy of the Nepal Heart Foundation was instrumental in securing government funding and support for a comprehensive control programme.[63] In India a National Rheumatic Heart Consortium was established in 2011 to formulate national guidelines and advocate to government.[64] Once programmes are established the Advisory Committee can offer valuable clinical, implementation and evaluation support.

Committee membership

A diverse Advisory Committee offers the best opportunity to address the primary needs of each stakeholder. Potential membership could include representatives from the categories outlined in table 2.

valuation oversight ngaging the community d other stakeholders	• Represent the programme to people living with RHD. Making it possible to tailor programme activities to best meet the needs of individuals, families and communities. • Track activities and data to guide the priorities of the control programme staff
Clinical leadership	• Develop evidence based, locally adapted clinical management guidelines • Develop consensus about management plans to standardise patient care and to provide clarity for clinicians at each level of the health system • Support excellence in care delivery
Representing the programme	• Present the work of the programme at meetings, in the media and to the community
Resource mobilisation	• Fundraise, mobilise resources or advocate for financial support of the programme
Advising and mentorship	• Support individuals and programmes to expand RHD control activities in other locations

> "From the very outset, it was understood that the success of local programs would ultimately depend on the extent to which countries integrated the program activities as part of their national health services"
>
> World Health Organization Report [65]

Terms of Reference

The goals of the committee should be clearly articulated in the **Terms of Reference (ToR)**.

The ToR, roles and activities of the committee may need to change over time as new priorities and challenges emerge. The ToR need 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. Many disease control programmes have an Advisory Committee and there may already be committees in your area with a ToR that you may be able to use as a model for drafting the ToR for the RF/RHD Advisory Committee.

Supporting autonomous programmes

New RHD control programmes are sometimes supported by international partners. These may include foreign 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. International support is generally provided for a fixed period of time. It is essential that these programmes develop a sustainability plan to continue the programme beyond the end of international support. Factors which supported early programme autonomy in the Pacific Islands include:[33]

- defined national strategy with feasible goals
- flexibility to amend policies
- regional and international collaborations and sharing of expertise
- clinical leader/champion
- stepwise approach
- integration with public health programmes over time

Things to consider

Do you have sufficient funding for your programme?

Does your programme have a fundraising strategy?

Do you have a business case or other evidence to support the need for increased funding?

How will you recognize 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 potential conflict of interest?

3. Funding

Securing long term funding for RHD control programmes is a global challenge. Worldwide, a number of registers, research programmes and other valuable efforts have been forced to close because of inadequate funding. [38, 66-68]

The clinical progression from GAS infection to RF and RHD can occur over of a number of years. This means that register based programmes are also needed for years or decades before burden of disease impact can be demonstrated (interim impact measures are outlined in Chapter 10). The best option is long term funding at predictable levels, although it may be difficult to convince governments to commit to multi-year funding for new programmes. In these cases, patching together multiple sources of shorter term funding may be necessary, with a plan to demonstrate value so that government can commit to longer term funding, ideally with a separate budget line item for RHD control.

Preparing for fundraising

A number of strategies for securing and maintaining funding may be helpful:

Making the case by estimating existing costs

Demonstrating over-investment in tertiary care can have a powerful influence on funding decisions for disease control programmes. Spending on admissions and heart surgeries often means that very little money is being spent on prevention.[69] Most of the direct cost of care for RHD occurs in hospitals and tertiary settings, so hospital admission and length of stay data (collected as part of burden of disease estimates) may inform cost of disease estimates.[70] For example, in New Zealand medical management of RHD accounted for 71% of the total costs of RF.[71] In Samoa and in Nepal the high costs of cardiac surgery helped governments decide to fund relatively low cost comprehensive control programmes.[63, 72, 73] Information from surgical waiting lists may signal a burden of severe disease which could have been prevented by the existence of a disease control programme.

Develop a draft plan

A programme plan with a draft budget demonstrates a readiness for action and may increase the likelihood that RHD might be included in a local budget allocation. Forming an Advisory Committee demonstrates the engagement of key stakeholders and can provide valuable planning input. Include estimated burden of disease data, the next steps you want to take and some provisional targets. Ensure the plan can be incorporated into your local integrated health plan, **NCD plan** or other process for decision making.

Develop a budget

A carefully developed budget will help potential funders have confidence in the programme and ensure that you are applying for the right amount of funding. Burden of disease data is helpful for estimating the number of people living with RF and RHD, which will also help you estimate staff, 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.

15, 78, 79

Individuals families and communities

Costs & socioeconomic burden of RHD

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.[74] Minimising the financial burden on individuals and identifying cost effective disease control strategies for populations is an important global goal.

"Sometimes, because of the patient's poverty, the cardiologist is obliged to choose among medications: instead of prescribing all the medication needed doctors have to prescribe just few drugs... We call this way of prescribing "managing prescription". Finally lack of finances causes the exacerbation of the disease and premature death."

Tchoumi et al, Cameroon, 2011.[80]

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 health care expenses; these 'out of pocket' costs drive medical poverty and personal bankruptcy.[75]

RF and RHD are costly due to reduced social and economic participation.[85] Young people with RF or RHD may be unable to complete schooling and parents may need to stay home from work to care for them. In Brazil, nearly one quarter of parents took time off work to attend to children with RHD and nearly 5% lost their job as a result.[9]

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.[76] In Egypt, 98% of parents of children living with RHD are concerned about the family and financial impacts of the disease.[77]

The health system cost of RF and RHD can be enormous, particularly in countries where governments subsidize or pay for health care. Most of the costs are incurred in tertiary treatment for severe disease, including hospitalization and surgery.

Indirect costs occur through the reduced economic participation of people living with RHD and of their families.

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.[5] There have been a number of projects to explore the cost effectiveness of RHD control in different settings.

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Some governments have directed international control programmes and humanitarian surgical missions, including USAID (Ethiopia[83]), AusAID (Tonga[81]), NZ AID (Vanuatu, Samoa[82]). Often these groups are arranged at a government level and require diplomatic consideration.[83] Northern Territory Community Organisations, faith based groups, business foundations and non-government organisations (NGOs) may provide valuable information for your programme.

Potential sources of funding

Many different kinds of organisations have funded RHD control programmes, equipment, events and activities. 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. Good luck!

Governments	In some countries, governments (local, state or national) provide a relatively reliable source of funding. Governments bear some of the cost of RHD- particularly paying for health care and reduced tax income when people are too unwell to work, providing an incentive to fund control programmes. Clinical advocates may be needed to access government budgeting procedures.
Professional Organisations	Professional groups, including medical associations or professional colleges, can be instrumental in generating or helping to secure funds supporting project activities. For example, funding from the American Legion to the American Heart Association was important for establishing community rheumatic fever programmes in the USA in the 1940s.[84]
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.
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.[86] 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 need 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, minimizing conflict of interest, reporting on outcomes and building financial sustainability is a specialty field in its own right. Larger programmes should consider reviewing fundraising resources or seeking expert advice.[76] Toolkits and resources include:

- Stop TB Partnership
- Heart Kids Hero

You may also be able to partner with fundraising or public relations organisations to develop a brand, strategy and fundraising materials for your programme.

Conflict of interest

Conflict of interest occurs when 'an individual or organization is involved in multiple interests, one of which could possibly corrupt the motivation for an act in the other'.[89] 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.

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, including tobacco companies and 'fast food' companies.

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, be recognised with a plaque or at 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.[81] 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. 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.

Case Study 2 | Tonga

Dr. Fakakovikaetau | Pediatrician | "Mafu Sai" Programme

The National ARF and RHD Prevention Programme in Tonga is formally recognized as part of the nation's National NCD Prevention Programme. An initial cross-sectional prevalence study of just over 5000 primary students performed in 2003 revealed the scale of the problem: a peak RHD prevalence of 11.5% was observed in children aged 10-15 years old. The "Mafu Sai" ("Good Heart") Programme was inaugurated in 2008 to address the high disease burden. Dr. Toa Fakakovikaetau, a paediatrician, shared her experiences:

Components of the programme

Funding for Mafu Sai is secured through the Ministry of Health and receives professional support from WHO as well as from New Zealand and Australia. The major focus of the programme is secondary prophylaxis. Echocardiography (echo) screening is carried out in two-week campaigns biannually. One local echocardiographer who was trained in 2010 attends the screening campaigns, in addition to technical support from New Zealand and Australia. Each year 3,000-5,000 children are screened via echo. In Year 6 of primary school, every student in the country receives an echo before they leave for secondary school as this is the highest prevalence age group. Many others are auscultated, however, and if a suspicious murmur is heard, children receive follow-up echocardiography. Children identified as having RHD are entered into a centralised register. Over 1,000 cases are registered to date with approximately 90% of cases classified as "mild" disease. Every child identified as having RHD, even if classified as mild and/or asymptomatic, is started on monthly BPG prophylaxis that is administered at their local health centre.

Open Heart International, a Sydney-based cardiac surgery team, visits Tonga every two years to perform surgical repair of valvular disease (most often RHD) as well as congenital heart disease. In 2013, 18 valvular surgeries were performed. With the aid of visiting cardiac surgeons, the need to refer patients for overseas surgery is decreasing significantly.

Challenges

Initially, there was little logistical knowledge of how to start a programme. Help from Australian experts and the RHD Pacific Working Group (2003) was instrumental in providing the necessary technical expertise and guidance. The 2003 prevalence study was instrumental in convincing the Ministry of Health to establish a national programme. The cost to the Ministry of Health of sending one patient abroad for surgery was equivalent to the entire annual budget for RHD screening and secondary prophylaxis efforts. In 2007/2008, programme facilitators experienced problems administering benzathine penicillin G (BPG). Health care workers reported that syringes were becoming blocked during administration. Quality of supply has since improved; there have been no further reported cases of blocked syringes, nor have there been quality concerns or procurement difficulties.

Moving Forward

Ongoing challenges include an understaffed local health workforce capable of conducting the screening campaigns independently. To combat the worker shortage, it is hoped that two echo technicians can be trained within the next 5 years. The programme aims to improve monitoring and evaluation capabilities. Local record-keeping and sharing of data is improving. It is commonplace now for staff at health centres to proudly show off their RHD statistics at meetings.

Tonga serves as an advocate for the creation of programmes similar in design to Mafu Sai in neighbouring island countries. It is believed that a focus on secondary prevention is especially effective in small island countries like Tonga.

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

4 Laboratory services

Time 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?

Access to laboratory services is a valuable component of RHD control programmes. However, successful programmes have been possible with very little laboratory support and it is not essential that complex laboratory services are perfected before disease control activities begin. The development of high quality laboratory services tends to be driven by the needs of many different diseases. Laboratory development can be expensive and requires policy makers and health system administrators to prioritise these aspects of disease control initiatives.[90] Advocacy to improve laboratory services can occur concurrently with other interventions to address RF and RHD.

Location and role of laboratories

Laboratory services parallel the levels of health care service (see page 10) – different laboratories fulfil different functions. Broadly, these can be considered local and reference laboratories, outlined in table 6.

Table 6: Laboratory roles and locations

Local laboratories	Reference laboratories
Local laboratories providing simple diagnostic tests for routine clinical use. Ideally, these 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.	Ideally a single national or regional streptococcal reference laboratory should be established.[20] Reference laboratories can provide critical support to local laboratories by: • Providing reference strains of GAS to ensure standardised results. • Providing expert advice on laboratory standards and training. • Providing specialist knowledge on sample testing and result interpretation. • Carrying out molecular typing. • Liaising with other national reference laboratories and public health bodies.

★ Opportunities for integration

The principles and practices of laboratory **bacteriology** are not specific to GAS. Establishing basic bacteriology facilities will greatly facilitate the clinical management of many patients with infections. Similarly, laboratory facilities capable of identifying GAS are valuable for more than RF alone; GAS causes a number of important clinical infections, and bacteriology facilities and techniques required for GAS isolation are applicable to a wide range of pathogens.

Evidence of preceding GAS infection

Throat swab cultures

Bacteria (including GAS) that are causing infection of the throat can be grown in the laboratory from bacterial swabs of the throat.

In order to isolate bacteria in the laboratory, the bacteria on the sample must be viable (i.e. still alive and reproducing). Bacterial viability can be adversely affected by antibiotic treatment of the patient prior to the sample being obtained. For many settings with a high burden of GAS disease, the use of antibiotics prior to presentation at health care facilities is common (See box 11, Chapter 13 – the role of chemists). Bacterial viability can also be adversely affected by delays in sample processing and exposure of samples to extremes of heat.

Asymptomatic GAS carriage in the nasopharynx also occurs, and may serve as a reservoir of infection. Results need to therefore be interpreted in the context of the patient's clinical presentation.

Tests for inflammation and/or infection

Rapid antigen detection tests

RADT tests are easy to use, commercial kits that detect specific parts of the GAS bacteria (antigens). There are a variety of kits produced by different manufacturers but the methods used are similar, and the advantages and limitations are broadly applicable. Sensitivity and specificity is variable between settings, kits and clinical disease.[92, 93]

GAS infection results in the production of **antibodies** specific to **antigens** expressed by GAS. 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 (ASO)** and **anti-deoxyribonuclease - B (ADB)** antibodies. Both tests are commercially available.

Acute phase reactants include ESR and C-reactive protein (CRP). There is variable elevation during the acute phase of the illness with arthritis or carditis. They may, however, be normal when **chorea** is the only manifestation.

Pre-operative evaluation

Laboratory tests for potential surgical candidates may include a full blood count, liver function test, creatinine, glucose, urea and electrolytes.[94] Screening for infectious diseases may include tuberculosis, HIV, hepatitis C and malaria. Women with RHD also require access to pregnancy tests. Details about preoperative evaluation appear in Chapter 23.

Anticoagulation

The **international normalised ratio (INR)** is a measure of therapeutic effect from the oral anticoagulant drug Warfarin. INR facilities are essential for programmes caring for people who have received mechanical heart valve replacement. Details about anticoagulation monitoring appear in Chapter 21.

GAS are divided into different types (strains) according to their *emm* typing. GAS have been separated on the basis of differences in the surface expressed M protein, however the sequence typing of the 5′ end of the M protein (emm) gene is now the preferred method for classifying strains of GAS.[95] Emm typing and genetic studies of GAS need to be conducted by specialist streptococcal laboratories. In general these are not clinically significant and are mainly used for research. Rarely, strain analysis may be a useful addition to outbreak investigation but this should be conducted in conjunction with a reference laboratory.[96, 97] Detailed laboratory studies of circulating community strains are likely to be needed for GAS vaccine development.[23] As progress towards a GAS vaccine continues, RHD control programmes may have a role in collecting this data – see Chapter 15 for programme engagement in vaccine development.

- ıd process management
- id customer focus
- Management of patient records and documentation
- recognition of outstanding staff.

Practical issues for laboratory services

Developing robust, reliable and quality assured laboratory services is a specialised technical field.[98] The details 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 in table 8 for your consideration.

Staff training

Formal training for laboratory staff will vary across the globe and may range from a short course to a degree from a university. 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 GAS to support remote training.[81] 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 consistent. Standards do not need to be complex, and have been established in a wide variety of settings despite variations in resources. Basic testing for GAS and for antibodies to infection require minimal resources.

Reporting of laboratory results

Samples which are being transported (even to local laboratories) will need to be labelled 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 clinical records.

Box 4: Sensitivity and specificity

Specificity

The specificity of a test is the proportion of people that are known not to have the disease who test negative for the disease.

Sensitivity

The sensitivity of a test is the proportion of people that are known to have the disease who test positive for the disease.

Organisation	• Structure of the laboratory, quality assurance responsibilities
Purchasing and inventory	• Resources and reagents to be purchased, received, inspected, stored and recorded.
Information and ocurrence management	• Identification and resolution of laboratory errors
Personnel and work environment	• Training of staff and work facilities.

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?

5. Integration with primary care & health systems

> "The implementation of the programme as part of the normal healthcare system's structure and facilities decreases the budget requirements and ensures the continuation of activities several years after the project ends"
>
> Nordet, Cuba, 2008.[26]

The integration of RF/RHD services into primary care services and other parts of the health system is widely recommended.[100-103] Integration is thought to be important for sustainability, quality of care and accessibility for patients and communities. However, integration is a difficult idea to describe, implement and deliver[104] and remains a challenge for RHD control programmes in most settings.[12, 105]

Integration with primary care

The foundation of primary health care is working at a 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.[106] The general principles of community based programmes transcend disease specific issues and focus on the needs of individuals and their families.[107] RF and RHD are ideally suited to a primary care approach; early signs (sore throat, joint pain) are often identified by primary care clinicians.[108,109] 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 is an important part of primary care and should be a core competency for front line health staff in most settings. Enhancing access to primary care is important for making primary care practical and affordable. For example, in New Zealand, six to eighteen year olds from high risk communities are offered 'walk-in' consultations with registered nurses for sore throat evaluation and treatment.[110]

Diagnosis 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 who receive the correct diagnosis of RF. Diagnosis relies on accurate use of the Jones Criteria (see Appendix B). In endemic settings primary care staff need sufficient training to recognise possible cases, and refer them for definitive diagnosis.

Delivery of secondary prophylaxis

Although register based programmes are helpful for ensuring consistency of BPG administration it may be possible to shift some secondary prophylaxis to primary care practitioners – particularly in rural or remote locations. Primary care systems may need adjustment to make this possible. For example, existing computer, **triage** or administrative systems can be adapted to include diagnostic codes and follow recalls for RF and RHD.[111]

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 with perinatal care

Women with RHD are at risk of heart failure and arrhythmias during pregnancy.[112] In late pregnancy blood volume increases by 30- 50% and heart rate rises.[113] During and after delivery blood pressure and cardiac output increase in association with contractions and changes in foetal demand.[113] These changes require considerable cardiovascular capacity and elasticity. In women with RHD this capacity may be limited by established heart failure or blood flow restricted by scarred rheumatic heart valves. Signs and symptoms of this cardiac deterioration are easily confused with late pregnancy: breathlessness, fatigue and swelling (**oedema**) and soft heart murmurs. Undiagnosed, cardiovascular collapse can lead to maternal death.

There is good evidence that RHD is a significant contributor to maternal mortality around the world. **Indirect obstetric deaths** are caused by underlying medical conditions made worse by pregnancy. [114] Indirect deaths account for 16.7% of maternal deaths in Africa and 12.5% in of maternal deaths in Asia.[115] In Egypt and South Africa a substantial number of these indirect maternal deaths are caused by RHD.[10, 116] In Saudi Arabia RHD was the underlying condition in 27% of pregnant patients admitted to the intensive care unit between 1994 and 2002.[117] In Fiji, RHD has been reported as the second most common cause of maternal death.[44]

Some women with RHD will become pregnant, others are diagnosed when they become symptomatic during pregnancy.[60, 67, 118] Some women will be undiagnosed until they become acutely unwell during delivery.

Although all deliveries with maternal heart disease are high risk it may be possible to minimise this risk by:

- Diagnosing the RHD prior to delivery and preferably prior to conception.
- Early antenatal care and planning for delivery.
- Delivery in medical facilities equipped to manage high risk pregnancies.
- Access to contraception.
- Although research is limited, strategies to improve integration with perinatal care could include:
- Educating all women with RHD that all pregnancies/ deliveries need close medical supervision, and providing tangible support for accessing medical care. Providing accurate and supportive education about RHD in pregnancy is very important; the social and cultural effects of having a condition which influences fertility may be significant.[11]
- Developing a referral system for primary health workers and midwives to access echocardiography and specialist review for women with rheumatic heart disease.
- Include RF/RHD in routine antenatal care, including medical history and cardiac auscultation.[119]
- Provide education for midwives about symptoms of heart failure which can be easily confused with the symptoms of late pregnancy.

Integration with chronic disease & NCD programmes

Policy

NCDs (defined by WHO as cardiovascular disease, diabetes, cancers and chronic respiratory diseases) caused 63% of deaths worldwide in 2008.[121] The burden of NCDs has continued to rise, prompting a High Level United Nations Meeting on NCDs in 2011. In 2013 all WHO Member States adopted the Global Action Plan on the Prevention and Control of NCDs (GAP).[122] The outcome of this meeting was an international commitment to reduce premature mortality from NCDs by 25% by 2025.[123]

Global momentum for NCD control provides a unique opportunity for RHD to be prioritised at a national level.[124] While it concentrates 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 rheumatic heart disease and other NCDs of childhood such as asthma, leukaemia and type II diabetes. In several countries where RHD is prevalent, health advocates have leveraged integration of RHD prevention and control into national health plans. For example, secondary prevention for RHD is included in the **WHO Package of Essential Non-Communicable Disease Interventions (PEN)** for primary care in low resource settings.[125] Similarly, national NCD plans offer an opportunity to integrate RHD prevention and control into national health plans.[126]

Practice

The sequelae of RHD, particularly heart failure, AF and stroke – are common non communicable diseases. These complications are often managed through NCD clinics. For example, in Rwanda, 25% of patients being managed with heart failure through an echocardiography service had RHD.[8] It is import that children and adults living with RHD can access services alongside people with ischaemic heart disease and other conditions.

Integration with congenital heart disease and other childhood heart disease

Congenital heart disease (CHD) shares some similarities with RHD- both affect young people, can be asymptomatic and lead to heart failure. Similiar medications are used for RHD and CHD and cardiac surgery is a component of management for both conditions. Screening programmes for RHD (see Chapter 20) will inevitably detect a proportion of children who have CHD and some will require interventions. It can be helpful to work with paediatric and CHD services to share resources (echo machines, visiting specialists) and provide support for families (children's heart clubs and heart disease education). For example, in Australia, the charitable group Heart Kids provides camps, advocacy and support for families living with a range of childhood heart diseases.[127] In India, a joint programme for diagnosis and treatment of RHD and CHD has been delivered through schools.[65]

Integration with healthy skin programmes

Although the association between group A streptococcal skin infection and RF remains unclear (see box 5) it is reasonable to consider including healthy skin programmes as part of RHD control.[16] At a clinical level there are a range of options:

- Risk factors for children and families with skin sores can be explored (particularly crowding) and the importance of treating infections (skin, throat and others) emphasised.
- In New Zealand some sore throat clinics (see Chapter 14) have expanded to deliver skin sore treatment programmes.[134] This programme aims to tackle untreated skin infections, the commonest cause of medical hospitalisation in high risk school age groups. The effect on GAS infections and RF is currently being explored.[135] This approach appeared to provide good access to antibiotic therapy, facilitated referrals for other health issues and increased **health literacy**.[134]
- Guidelines for management of skin infections may be added to training or education material for sore throat treatment, taking swabs for culture or laboratory protocols.[136]

Clinical integration may be paralleled by advocacy, policy and research collaborations for the control of skin diseases.[137] For example, efforts to reduce household overcrowding are likely to reduce the burden of both skin infection and RF.

Integration with dental care

RHD increases the risk of **infective endocarditis (IE)**, a serious bacterial infection of the heart valves. The bacteria that cause IE are often found in the mouth and excellent dental hygiene is needed to keep teeth and gums healthy and to reduce the risk of IE. Although there is a clear link between good dental care and RHD outcomes there have been few attempts to integrate these services.[138] In New Zealand, young people with RHD received toothbrushes and toothpaste from district nursing services.[139] Other opportunities for integration could include:

- Including dental representatives on your advisory committee (see Chapter 2).
- Having a relationship with a dentist working near your hospital or clinic and physically connecting people with RHD to dental services.
- Included dental care on your priority based care plan pathways (see Chapter 20).

Things to consider

Does your programme provide a clear consistent message about RHD control to local, regional and national governments?

Do you have resources available to ensure that all advocacy activities are 'on the same page' 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?

6. Government engagement & advocacy

Governments are one of the most important stakeholders in RHD control.[20] Often they are responsible for overseeing the health and education systems critical for the prevention and treatment of the disease. Government policies also have a significant impact on the primordial determinants of disease. These important roles make government engagement in RHD programmes essential.[72, 140] However, engaging government often depends on local custom, politics and government structure. Approaches to government are best guided by experience in the local setting

Identifying specific 'asks'

Effective advocacy requires a clear vision of what you are asking for and why you need it. 'Asks' will depend on your local situation, priority setting by your RF/RHD Advisory Committee and the stepwise conceptual framework outlined on page 6. Some of the preparatory work for fundraising (Chapter 5), including burden of disease estimates and a plan for intervention may also be helpful.

Advocacy requests for RHD control may be broad: including RHD on national strategies, adding RHD to the agenda of local meetings or ensuring there is protected time for health staff to attend specific training. Higher level requests may be appropriate in some settings, including workforce planning, resource commitments to achieve consensus goals and prioritizing RHD in the national agenda. See also the clinical advocacy section on page 34. An example of a clear ask is the Paediatric Cardiac Society of South Africa publication: Optimal paediatric cardiac services in South Africa – what do we need?[141]

Clinical advocacy

Clinicians can be powerful advocates for RHD control. This may include professional groups or the work of individual clinical champions as outlined in table 9.

The World Heart Federation has an Advocacy Toolkit available online to assist with building public support for health needs.[146] The toolkit outlines nine key steps to effective advocacy outlined in table 10.

Clinical organisations have a credible professional voice to call for resources, attention and action to RHD. This can have a powerful effect on governments.

For example, a concerned group of cardiologists and cardiac surgeons committed to support efforts in a statement known as the "Drakensberg Declaration" 2005 , updated in 2011.[142]

In Pacific Island countries clinicians developed a "Call for Action" from a workshop held in Fiji in 2006, and again endorsed at a subsequent workshop held in Fiji in 2008.[143] 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.[143]

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 New Zealand College of Public Health Medicine has a policy statement on RF, supporting government investments in a throat swabbing programme.[145]

'RHD champions' have been critical disease advocates in a number of settings.[5] Worldwide, clinical advocates have been critical for maintaining RHD on the national and international agenda.

For example, Professor Bongani Mayosi, South African cardiologist and researcher has been a vocal advocate for RHD control.[145]

'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 health care started to turn.'"[145]

Community engagement

In most 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. Community initiated projects, such as the Papago RHD control project in Arizona[147] have demonstrated the feasibility of engaging high risk communities in comprehensive control programmes. Petitions, calls to action or community consultation may provide additional opportunities for engagement.

Government engagement

Governments are the most appropriate group to support, fund and oversee RHD control programmes over the decades required to achieve disease control. This may include roles for the Ministry of Health, the Ministry of Education and other ministries responsible for housing and economic development.

Opportunities for integration may include:

- Aligning RHD control programme priorities with national health plans
- Development of national RHD strategies
- Working towards common goals for disease control
- Undertaking shared research projects
- Providing local data to support decision makers
- Engaging with local/regional public health strategies

★ Opportunities for Primordial Prevention

RHD is a disease of poverty and is rare in economically developed settings. Governments are the major determinants of social and economic policy to address the conditions of poverty, overcrowding and inequality which contribute to RF and RHD. RF has declined where governments and their population have improved environmental conditions and provided access to resources that improve hygiene.[154] Educating communities is a core component of an RHD programme. Programmes can assist government through the provision of useful data, and by highlighting the importance of improved housing and health care services.

Engagement with the Ministry of Education

"The school occupies a unique position in relation to rheumatic fever control."
American Academy of Pediatrics, 1948.[155]

School age children are at greatest risk of RF, making schools a valuable 'front line' 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. 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 new student transfers may be possible.[149] Delivering education and services through schools generally requires support from the Ministry of Education, and often at an individual school level.[150]

A range of education integration models 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. [25]
- In South Africa school health nurses are being trained to provide antibiotics for children with sore throats.[151]
- In New Zealand diagnosis and management of sore throats and skin infections is integrated into schools in high risk communities.[135] (See Chapter 14)
- In India the Rupnaga District Project is training school teachers to identify suspected cases of RF.[64]
- In Samoa collaboration with the Ministry of Education, Sports and Culture was a significant aspect of a pilot programme implemented in selected schools.[152]

Other opportunities for integration may include:

- Training teachers to provide education on RF/RHD to students (See Chapter 11).
- Train teachers to identify symptoms of joint pain and abnormal movements (potentially chorea, see Annex B) and encourage parents to seek medical assistance.[153]
- Deliver some health services through schools, including school nurses and RHD outreach activities (See Chapters 14 and 20).

Case Study 3 | Nepal

Thomas Pilgrim, MD | Assistant Professor
Department of Cardiology Swiss Cardiovascular Center, Bern University Hospital, Switzerland

"Our involvement in the initiation of an interventional cardiology programme in Eastern Nepal brought our attention to the high burden of rheumatic valvular disease in that part of the world, and stimulated our interest to extend our focus to timely prevention of subclinical rheumatic heart disease in children."

The collaboration with B.P. Koirala Institute of Health Sciences (BPKIHS) started with Dr. Pilgrim's support in establishing the first cardiac catheterisation laboratory in Eastern Nepal. Taken aback by the high prevalence rates of advanced valvular disease in the young, and at the same time realising the limited access to health care for less privileged patients, Dr Pilgrim's team decided to complement the programme with prevention component targeting RHD. The team began to screen children from governmental and non-governmental schools in rural and urban regions of the Sunsari district, situated on the foothills of the lower Himalayan range, in 2012.

Components of the programme

The programme designed a study protocol to investigate the prevalence rate of silent RHD among schoolchildren in the Sunsari district in Eastern Nepal and aimed to determine risk factors for progression to manifest disease, evaluate barriers for receiving adequate care and assess long-term clinical outcome. The study protocol has been approved by the local institutional review board and the Nepal Health Research Council.

"Education is the first priority."

The project was broadcast in local radio programmes and newspapers in order to raise awareness. A 15 minutes orientation video has been produced and is screened prior to the initiation of the screening examinations at schools for the information of children and parents. The short movie narrated in Nepali gives a concise overview on the nature of RF and RHD and explains the rationale for screening and stresses the importance of secondary prevention.

"Communication and orientation through local key opinion leaders."

The programme tried to win local key opinion leaders as advocates of the programme: officials from the district education office, school principals, local health care professionals and parents. Active involvement of local people rooted in the community will increase sustainability of the programme and strengthen established structures.

"We built a team"

Screening campaigns in 12 schools have been performed since the beginning of 2013 by a dedicated team of Nepali health care professional. After a short medical history and physical examination onsite screening is being performed using a portable ultrasound device. Children with findings consistent with borderline or definite RHD are invited for confirmation and initiation of secondary prevention to the outpatient clinic of BPKIHS in Dharan. Regular clinical follow-up is performed at BPKIHS.

"Early detection of silent disease"

Early detection of silent disease and timely implementation of secondary prophylaxis may prevent progression to clinically manifest disease. Children who screened positive for valvular lesions consistent with RHD are educated together with their families about the importance of secondary prevention and regular clinical follow-up. Secondary prophylaxis with BPG is provided by the Nepal Heart Foundation and administered at B.P. Koirala Institute of Health Sciences. A centralised registry has been developed to support and monitor patient compliance and clinical follow-up.

Challenges

The programme encountered several challenges in the run-in phase of the programme.

Stigmatisation by subclinical disease

It was often challenging to explain the significance of subclinical disease and stress the importance of secondary prevention in asymptomatic children to parents and primary caregivers. Programme implementers tried to anticipate anxiety related to screening and potentially stigmatising findings beforehand by education and offer continued support to children identified with heart disease.

Incidental findings

During echocardiographic screening several children were diagnosed with clinically relevant congenital heart disease requiring medical attention. Adequate cardiac care is offered for incidental findings detected during echocardiographic screening.

Collaboration with primary care physicians

Certain children identified with subclinical rheumatic heart disease and included into the longitudinal registry were recommended to cease secondary prophylaxis by their family's primary care physician. Inconsistent medical advice by different health care providers decreases the motivation to adhere to regular secondary prevention. Outreach and involvement of primary care physicians with an established relationship with the families is of key importance in order to guarantee long-term adherence to secondary prevention.

Are there notifiable diseases in the setting?

Are RF or RHD notifiable?

How do you define suspected or confirmed cases?

If notifiable, can notifications be automatically added to the RHD register?

7. Disease notification

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.[156] Making a disease legally 'notifiable' by doctors and health professionals allows for intervention to control the spread of highly infectious diseases such as influenza, poliomyelitis or yellow fever. In less infectious conditions it improves information about the burden and distribution of disease.

In endemic settings RF – and potentially the first diagnosis of RHD- meets the broad criteria for suitability as a notifiable disease, outlined in table 11.[157]

RF has been made notifiable in many places: New Zealand,[63] Australia,[28, 158] Fiji,[159] South Africa,[160] New Caledonia,[161] Tuvalu, Kiribati and the Solomon Islands.[162] In Fiji RF is one of 46 diseases notifiable each week by all medical practitioners.[159] However, clinician education and engagement limits participation; only 43% of government health facilities provided a notifiable disease report in 2000.[159]

In South Africa RF, and the first diagnosis of RHD, were made notifiable in 1989. However, by 1992 the initial diagnosis of RHD had been removed from the notifiable disease list.[160, 163] Although amended, there appears to be significant under reporting of RF notifications.[160] Of concern, nationally reported figures in this study implied a decreasing burden of RF, despite a consensus opinion by cardiologists of an 'RF Epidemic'.[160] This illustrates the risk of incompletely implemented RF notifications.

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 required.

Clinicians often have considerable demands on their time and may be unfamiliar with reporting requirements. Significant underreporting is common. Clear guidelines are helpful for communicating and disseminating case definitions and pathways for reporting. The New Zealand 'Communicable Disease Control Manual' is a good example of this approach – the RF chapter includes: case definition, case classification, notification procedure, case management and contact management.[164]

A preventable disease	RF appears to be preventable at a population level by changing living circumstances (see Chapter 26). High quality secondary prophylaxis can be disease altering following an episode of RF, prompting calls for RF to be made a notifiable disease.[157]
There must be an identified population or sub-population targets	Young people at greatest risk of RF often come from low resource or vulnerable communities.

Closing the feedback loop

One of the barriers to clinicians reporting RF or RHD can be a perception that the data is endlessly gathered but not used to make changes.[159] 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.

★ Opportunities for integration

In countries without strong notifiable disease programmes, clinicians with an interest in RF/RHD may work with other potentially notifiable diseases (for example: tuberculosis, leprosy, dengue and many others) to develop reporting systems.

"We cannot begin the long journey [of disease control] until we make [RF/RHD] reportable."

Christie, California, USA, 1941.[155]

things to consider

- Does your programme have an RHD person – a key individual already working in RHD or interested in the area, who can take over programme coordination responsibilities?
- 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 shortage in your area?

‘The program manager, can liaise effectively with schools, school health services, primary health care centers and maternal and child health services as well as with departments of medicine and paediatrics and laboratory services in hospitals’

Dodu & Bohig, 1989.[62]

Human resources

All RHD control programmes require staff to help run the programme and deliver care. However, 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. Identifying one key person to coordinate this work is critical. Wherever possible, these key individuals should be supported by a number of clinical and non-clinical staff.[165]

Allied Health	• Laboratory staff • Epidemiology support

Box 6: The importance of 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.[63, 67]

Case study: Kiribati[43]

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, educational materials were 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.

Whilst 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. At the end of the current externally funded programme the position in Kiribati will be continued by the Ministry of Health.

emuneration

Differential between source and destination country can be a significant motivator to leave, and barrier to return.

Specialist clinical human resources

Paediatric cardiology

The global shortage of specialist clinicians is particularly acute in areas where RHD is endemic.[166] 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.[168]

Paediatric cardiothoracic surgery

Few paediatric cardiothoracic surgeons are available in low and middle income settings. Sub-specialty in developing countries is quite low. Using India as an example, in 2005 it was reported that there were only 10-12 paediatric cardiac surgeons. "a small fraction of what is required for optimum care".[168] Sustainable funding to operate training programmes is difficult in environments where finances are limited. Some authors suggest that only large scale political and socioeconomic change will see developing nations realise change that provides greater access to paediatric cardiothoracic surgery.[169] Increased awareness amongst existing cardiologists, sharing of adult cardiology resources, and specialty colleges taking the lead in training programmes have been suggested as measures that can gradually increase this valuable resource.[168]

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 echo 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.[170]

★ Opportunities for integration

Echocardiographers may have a role in reporting newly diagnosed RF and ensuring that the clinical status is updated in registers.

Nurses and midwives

Health systems across the globe grapple with a critical shortage of nursing staff:

- Sub-Saharan Africa 11 nursing/midwifery personnel per 10,000 population versus 49 nursing/midwifery personnel per 10,000 for the Americas and 78 nursing/midwifery personnel for Europe.[172]
- Globally, high income nations average 87 nursing/midwifery personnel per 10,000 population versus 11 nursing/midwifery personnel per 10,000 population in low income nations.[172]
- Wages, education, training and access to medications, safe water and essential medical equipment are all areas of priority in rectifying the global nurse shortage.[173]

Strategies for retaining health workers

"Retaining health workers can be done by providing adequate salary, something as simple as adequate and safe working equipment, clear job descriptions and at least some form of continuing education"

Leblanc, 2009.[174]

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.

Administrative strategies

The global health workforce crisis makes it difficult to retain health staff, particularly when they 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. Many factors contribute to health worker migration by encouraging departure (push) or encouraging recruitment to a new setting (pull), some of these factors are outlined in table 13.

Employment opportunities	Availability of jobs, job security during times of budget cuts and public service retrenchment.
Working environment	Excessive workloads, poor working conditions, low staffing levels. Human resource systems that are inadequate for the environment.

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, tele-health or online approaches to education and training materials

How can training material be evaluated and improved?

Informed and engaged health workers are a critical component of successful disease control programmes. Without training, guidelines will not be used, patients will get inconsistent messages and follow up may not be delivered to those most in need. However, providing training can be difficult in settings with many competing health priorities.

Develop an education and training plan

An important part of RHD control programmes is the training and education of staff and affiliated health workers. RHD control programmes should support all health staff to improve knowledge, expertise and skills in the prevention, diagnosis and management of RF and RHD. Education, training and the dissemination of information increase capacity and improve outcomes.[158] A plan for training activities, expected competencies and outcomes should be developed. Training does not need to be elaborate. In Mali, a single day of teaching on clinical dermatology produced significant improvement in quality of care and cost reduction. Improvements persisted for 18 months.[176] Providing primary health care staff with basic training to identify suspected cases of RF and RHD offers the best chance for identifying early disease and beginning intervention.[177]

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 areas - particularly the geographically dispersed Pacific Islands - for improving management.[178,179] owever, 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. A number of curriculums for different types of RHD training courses have been developed, outlined in box 7.

Ensuring that RF and RHD are included in existing local training materials for health worker, nursing, midwifery and medical training is an integrated and relatively low cost intervention. However, 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 working in high risk settings is a valuable way to improve diagnosis and management.

Box 7: Sample training resources

World Heart Federation Curriculum[19]
The WHF curriculum provides a brief overview of core requirements in RHD and is available via RHDnet in a number of languages.

RHD Australia Online Modules[28]
A suite of on-line modules for clinicians to develop 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.

Ethiopian Health Centre Team Module[81]
Formal training modules have been developed in Ethiopia as part of the Public Health Training Initiative, to educate health care 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.

> “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.”
>
> Chicago Rheumatic Fever Program, United States of America, 1982.[182]

Publications

It may not always be possible to bring health workers together to learn about RHD. In these cases, publications, mail outs, posters or other resources can be used to share information for health professionals. A overview of this approach appears in table 15.

ox 8: High staff turnover

ommunities with a high burden of RF and RHD so tend to be areas with a high turnover of health orkers.[67] This makes ongoing training- and the evelopment of accessible resources- particularly nportant. Training and materials should cover essential inical knowledge and the main principles of how your RHD ontrol programme works.

n particular, new staff should be made aware of the ymptoms of RF and the need for specialist evaluation o confirm diagnosis. Every episode of RF which goes nrecognised is a missed opportunity to begin life hanging secondary prophylaxis.

ducation on how to notify cases, referrals and nterventions may also be needed. Orientation for new aff is particularly important if they have come from igh resource settings where RF is rare.[110]

★ **Opportunities for integration**

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

Table 15: Publications including RF and RHD

Dedicated RF / RHD publications	Embed in existing publicationns
The World Heart Federation website for RHD, RHDnet, is an international resource developed primarily for clinicians, health practitioners and policy-makers in countries where the disease is still common. Its purpose is to promote RHD control through best practice including registration of people with the disease and secondary prevention of RF: Loccal guidelines/handbooks for the management of RF and RHD can be developed. For example, the RHDAustralia website contains an online clearing house of resources, newsletters and blog articles.	An ever-increasing number of clinical guidelines and algorithms can sometimes become overwhelming for clinical staff. Ensuring that local guidelines are incorporated into existing standard resources minimises this problem. For example: • The Integrated Management of Childhood Illness (IMCI) programme in Africa includes sore throat guidelines. • The popular handbook for people without medical training in remote locations, Where There Is No Doctor, includes RF.[180] • In Rwanda, managment of RHD is outlined in detail in the PIH Guide to Chornic Care Integration for Endemic Non-Communicable Diseases.[181] • The CARPA Manual from Australia provides information for health professionals about diagnosis and management of RF. • The WHO Pocket Book of Hospital Care for Children includes information about management of RHD.

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?

10. Programme evaluation

Monitoring and evaluating the success of your 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

Monitoring involves continuous checking of the programme to ensure that it is proceeding according to plan. Ask the question "Is our programme progressing in such a way that its goal will be achieved?"

Monitoring is conducted by collecting data (indicators) at regular intervals (monthly or yearly) to measure the extent to which:

- Programme activities are taking place (process indicators)
- Programme objectives are being met (outcome indicators)
- The programme goal is being achieved (impact indicators)

Evaluation involves determining the relevance, adequacy, effectiveness, efficiency and impact of programme components. Different types of evaluation can be undertaken at different stages of the programme. A formative evaluation can be carried out during the planning phase, a process evaluation during the implementation phase, and a summative evaluation at the end of the programme.

Depending on the approach, evaluation asks the question, "What is the best way to achieve our goal?", "Could our programme work better to achieve our goal?" or "Could our programme have worked better to achieve our goal?"[183]

Approaches to evaluation

Narrative review

Historically, evaluation of RHD control programmes has been in narrative form, outlining the project and describing outcomes. Often these reports are required by donor agencies or other funding groups. This approach has identified a number of valuable lessons in South Africa, Kenya and the Top End of Australia.[184-186] Narrative reports are usually free text which can make it difficult to compare numbers or progress over time.

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".[187,188] Clinical audits in low resource settings are a valuable opportunity to improve quality of care, though there are multiple barriers to undertaking audits and changing practice.[189, 190] Particular challenges may include: unclear audit criteria, absence of guidelines, fragmented health systems and limited human/financial resources.[190]

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. Audits have been completed for a number of RHD control programmes.[66, 191,192] Surgical outcomes require a specialised clinical audit approach outlined in Chapter 24.

Continuous quality improvement

Improving the quality of health care delivery is an essential consideration for low resource settings.[193] **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 cycle, although it 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 cyclic, therefore measures to improve practice can be implemented and evaluated, and as part of the process the team members integral to improvement increase their knowledge by participating.[194] An extensive package of CQI interventions for RHD programmes has

been developed in the Northern Territory of Australia.[195, 196] In this setting 'CQI also provides a structure to refine and reinvigorate programmes to promote sustainability.'[197]

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 best practice guidelines to use as CQI benchmarks.[28] 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.[158]

Contracted or external monitoring and evaluation

External evaluation can provide important and independent assessment of how your programme functions.[198] The Advisory Committee can develop Terms of Reference for review to ensure that the evaluation addresses priority areas. Evaluation resources can provide a broad overview of the role, benefits and expectations of monitoring and evaluation.

★ Opportunities for integration

Could you work with a nearby programme to complete peer programme evaluation?

Box 9: Experiences of people living with RHD

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.[199] As many people living with RHD are young, appropriate ways to obtain their views should be utilised, for instance, drawing what RHD means to them. Ensure interviews are conducted in the main language spoken. Include some open questions to add depth to the evaluation.

Case Study 4 | Ethiopia

Professor Abraham Haileamlak | Paediatric Cardiologist | Jimma University Hospital

A formal national RHD prevention programme does not exist in Ethiopia, but a taskforce of physicians from medical schools in each corner of the country work together to advocate for RHD prevention, up to the level of the Ministry of Health. One of these physicians, Dr. Haileamlak, a paediatric cardiologist at Jimma University Hospital, reflected on his experiences instituting a school-based screening programme in south-western Ethiopia, from February 2012 to June 2013.

"We reached our target."

The project—while it does have a service delivery component for those children identified as having RHD by screening—was primarily a research project funded by Elsie Kröner-Fresenius-Stiftung with a goal to establish a baseline prevalence of RHD in south-western Ethiopia. The project reached its goal of screening 2,000 children in 17 schools. A prevalence of 27/1000 was calculated, with 11 cases of borderline RHD also identified. A central register was created to keep track of the RHD positive patients.

Components of the Programme

"My phone rang with questions from parents."

Professor Haileamlak considered this as evidence of a successful awareness campaign. Education about RHD was provided to teachers on the day of echo screening in each school. Students and their families were often educated only in the case of a positive screen for RHD. Broader awareness campaigns existed in the region, often in the form of a radio broadcast that was unrelated to this screening project.

Echocardiography screening in schools was conducted by a mid-career health professional. A second opinion from Dr. Haileamlak was sought in questionable cases. Nurses recorded patient information and performed vital measurements. Children identified as having RHD then visited the Jimma University Hospital for follow-up and further education with their families. Secondary prophylaxis with benzathine penicillin G was initiated at this visit. Subsequent dispensing of secondary prophylaxis was carried out in local health clinics at a cost to patients and their families. Patients were seen at Jimma University Hospital every 4-6 months for follow-up. Compliance with secondary prophylaxis was not formally documented, although questions to assess compliance were asked of patients at each hospital visit. It is too early to assess patient adherence or long-term effects of the programme.

Challenges

"Many donors are not interested in RHD."

The project was constrained by funding. Fuel for transport to school screening sites, over 50km one way, was a significant unmet expense. In addition, distance to the schools posed a cumbersome time commitment.

"There was resistance in certain schools."

Resistance usually came from teachers who sought an incentive in exchange for allowing their pupils to be screened. Prior to education, some resistance from families and students occurred. Finding an echo technician was initially difficult. In retrospect, Dr. Haileamlak believes it would have been more efficient to carry out the screening on his own in order to reduce the need for repeat echocardiography. Because only a random sample, and not all of the students at the 17 schools were screened, issues arose when parents whose children were not selected for study also wanted their children screened. To remedy this, children outside of the research project were often screened but their results were not included in the data. It is unclear what follow-up these children were offered. No surgical option existed for patients with severe lesions, but there are plans to have surgical facilities in place within the next 2-3 years.

Primary Prevention

Primary prevention encompasses interventions to prevent the development of RF. Typically, this has entailed treatment of GAS infections in young people. Prompt treatment of GAS 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 GAS vaccine has the potential to revolutionise primary prevention by preventing GAS infection and subsequent development of RF. A vigorous research agenda to develop a GAS 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 GAS 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.

Things to consider

Who are your priority target groups?

What do you want them to do?

What do they need to know, to do it?

What is the key message for your targeted priority group?

How will you test messaging to ensure it make sense to the target audience?

Who can help you develop materials?

Do any of your collaborators already have experience communicating with your target audience?

How will you evaluate the impact and reach of your communication?

How will you sustain your communication with target groups over time?

How do you record media coverage about RHD?

11. Community education

Communities, families and individuals are critical stakeholders in the control of RF and RHD. RHD prevention requires an engaged community that knows when and how to seek care.[22, 61] However, relationship of GAS 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.[200]

Linking sore throats to joint pains and RHD is a critical component of community education. There is no 'right way' to develop an education programme; this chapter aims to provide some guidance while further research is underway.[201]

WHO has identified six components of successful community health awareness campaigns.[202] These components are outlined in table 16 with reference to RF/ RHD specific education.

'Talking Wall' developed by the Kenyan Heart National Foundation to raise awareness of RF.

	Existing community resources and infrastructure should be identified and evaluated to provide the local needs. For example, populations will require a different approach, reflecting their existing knowledge and information 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, and the goals of the campaign established. Clear messages reduce confusion and improve retention.[202] Examples include: • "Rheumatic fever licks the joints and bites the heart" • "Sore throats make sick hearts" (Fiji[179]) • "Sore throats can lead to a broken heart" • "Say Ahh Campaign" (New Zealand, 2013) • "Sore throats matter"
Comprehensive programme	Comprehensive RHD control programmes address community education at a variety of levels; including children, parents, teachers 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 epicenter for RF/RHD prevention should not be solely in the health clinic, but at schools, homes, faith based centers and community spaces.
Long-term change	A successful programme will establish a sturdy foundation that can serve as a platform for enduring change. Education should be established to promote sustainability, rather than occur as a "one-and-done" outreach effort.

★ Opportunities for integration

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 maximize outcomes. You may be able to access support through hospitals, health departments, businesses, universities or charitable groups.

Media options

Pamphlets

Pamphlets 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 taken home from school.[204] 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.[63, 203] In Kenya, the Kenyan Heart Foundation ran a programme to paint thirty two 'Talking Walls' in schools, displaying signs and symptoms of RF (illustrated on page 43).[186]

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 television advertisements. In 1996 an RF/RHD information video was produced for the Northern Territory of Australia, funded by charitable donations.[85] 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 education tool.[85] Producing videos can be time consuming and expensive, so it is important to have a clear vision for how audio-visual material will used and where it can be distributed. Interviewing or involving policy makers in the film may be a way to engage their interest in RHD control. A number of educational video resources are available online:

- Sore throats can lead to a broken heart
  South Africa
  As part of the STOP RHD ASAP Programme in South Africa, students have created a six minute film in which four adolescents with RHD are featured. Stories are a critical component of community education: "while professionals learn through data, communities learn through stories"[202]
- Information about rheumatic heart disease
  Australia
  A 10 minute video featuring local actors, musicians and locations in Queensland, Australia.
- Nepal RHD video
  Nepal
  A 15 minute video in Nepali with English subtitles about prevention for RHD.

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 module for RHD has been piloted in Kenya.[205] In this module, animated presentations link sore throat to RF and then RHD. Immediately after completing the module, students answer interactive questions with prompt feedback. Anecdotal feedback has been very positive and formal evaluation is underway.

Mobile phone text messages have been used in some areas to provide general public health messages, although not yet explored in the context of rheumatic heart disease.

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.[200] A number of toolkits and social media resources are available online.

Radio and television

Radio messaging may be particularly useful for dispersed populations or in times of social instability and in areas of low literacy.[206] Radio messaging has been used in New Zealand,[207] South Africa,[24] Nepal,[64] and the French Caribbean.[26] Programme evaluation from New Zealand found radio messages had been well retained by the target audience.[207] Radio messages could include:

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

Media training for doctors or others who are going to be interviewed on radio or television can be helpful.

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.[208] In New Zealand a 'Colouring In' competition was developed for 5 – 6 year old children to illustrate a picture of a sore throat and reinforce the message to tell an adult about throat pain.[209]

Celebrity endorsement

Celebrities can provide a powerful message about the importance of RHD control.[168] 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.[210]

Events

A wide range of events are possible to increase awareness. The annual 'Rheumatic Fever Week' is held in August in South Africa each year.[24] A Heart Club founded in Kerala, India hosts events for young people living with RHD.[210]

Education for specific audiences

School Students

School-based education is an effective strategy to target young people most at risk of RF and RHF.

Education can be incorporated into regular activities:

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

Teaching should also occur alongside events dedicated to RHD control, including echo screening or sore throat swabbing 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.[204] However, engaging teachers with the programme appears to be important for the transmission of knowledge from teachers to students.[204]

In Sudan, teachers receive a manual about RHD, and hundreds have seen a video about RHD as part of their training.[211] 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.[26] In Nepal, students have written poems, performed skits and sung songs about RF/RHD to raise awareness.[63] 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.

★ 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.[200] 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.[208]

Education for people newly diagnosed with RF or RHD

People living with RHD need even more information about the disease. However, without a strategy for consistent communication and education many remain confused about their own condition.[200, 212] Approaches should include:

- Culturally appropriate conversations with clinical staff who ideally speak the same language.
- Information resources which can be easily understood, even in areas of low literacy or multiple languages.
- Advice about concerning symptoms, or when to seek help.
- Advice about dental hygiene and prevention of infectious endocarditis.[138]
- Supportive advice about contraception and careful management of pregnancies.

People with a new diagnosis of RHD should also be referred any local **support groups**, particularly rheumatic heart clubs.

12. Severe and Rheumatic Heart Disease

Sore throat (pharyngitis) is a common childhood disease in most countries. In settings with endemic RF/RHD approximately two thirds of these sore throats are caused by viral infections. One third of sore throats are caused by a bacterial infection, most commonly group A streptococci (GAS).[213, 214] Treatment with oral penicillin can reduce the attack rate of RF following GAS by about 70% and up to 80% with IM penicillin.[215] A full 10 day course of appropriate antibiotic treatment started within nine days of sore throat symptoms can prevent almost all cases of RF.[215-217] In settings where RF and RHD are still common, diagnosis and antibiotic treatment of streptococcal pharyngitis are critical, but in countries where RF and RHD are rare, there is an increasing tendency to recommend against antibiotic treatment.[218]

Distinguishing viral pharyngitis from bacterial pharyngitis is difficult. A wide range of clinical guidelines have been published by organisations in high resource settings.[218, 219] 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.[220] In addition, the clinical presentation of sore throat varies significantly between low income settings.[221] Developing local guidelines for the diagnosis and treatment of sore throat is an important role for RF/RHD control programmes.

Why develop (or adapt) local clinical guidelines?

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.
- Deliver care which meets the needs of local communities.

Treatment approaches

Different programmes and places will make different choices about GAS treatment guidelines. These tend to reflect local opinion, experience and resources.[222] Factors to consider include: identifying the greatest proportion of GAS infection, minimising costs of diagnostic tests, minimising unwarranted use of antibiotics.[15] Broadly, there are three main approaches to managing symptomatic childhood sore throats in endemic settings, outlined in table 17.

Clinical scoring tool + treat	A range of clinical scoring tools have been developed to try and distinguish GAS pharyngitis from viral pharyngitis.[181, 226, 227] These appear to have some use in identifying sore throats which are most likely to be caused by GAS. However, scoring tools tend to be highly sensitive (correctly identifying children with GAS pharyngitis) but poorly specific (incorrectly identifying children with viral infection as having GAS pharyngitis). This means that many children will receive unnecessary antibiotics. (See box 4, Chapter 4 for an explanation of sensitivity and specificity).

octors are sometimes worried that treating too
any sore throats with antibiotics will cause antibiotic
sistance.[1,215] Use of penicillin to treat sore throats has
ot been associated with penicillin resistance in GAS; no
AS isolate has ever demonstrated penicillin resistance.
ne mechanism of this persistent sensitivity to penicillin
poorly understood but has been maintained for many
ecades with widespread use.[228] There is the potential
r over-use of antibiotics for pharyngitis to contribute
resistance in other bacteria, particularly if penicillin
amoxicillin were replaced by broader-spectrum often
ore expensive antibiotics. For example, Streptococcus
neumonia is a major cause of pneumonia which
as demonstrated resistance to penicillin following
idespread overuse of antibiotics to treat for viral
fection.[206] It is important that systems to support
tional prescribing and accurate diagnosis of GAS are in
lace and updated regularly. The use of broad spectrum
ntibiotics for pharyngitis should be discouraged.

Antibiotic selection

Penicillin for primary prevention of RF can be administered orally or intramuscularly (IM). Adherence is also improved with one-time BPG injection because strict compliance with a twice-daily oral regimen for 10 days is difficult to maintain for many patients.[229] Erythromycin is suggested as an alternative for patients with a proven history of hypersensitivity reaction to penicillin.[29]

If your guidelines include oral antibiotic options it be may worthwhile to consider how individuals and families can be supported to complete the full course of medication.[232] Strategies may include:

- Provide a clear verbal explanation to continue antibiotics, even if symptoms resolve.[134]
- 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.[209]

Table 18: merits of oral and injectable antibiotics for primary prophylaxis

	Advantages	Disadvantages
Oral antibiotics (generally 10 days of penicillin V or amoxicillin)	Minimal discomfort	Adherence is usually poor.[230] Paediatric formulations may require refrigeration.[231]
Intramuscular injections (benzathine penicillin G)	Guaranteed adherence	Painful. Possibility of adverse drug reaction

things 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."

Grigg et al, New Zealand, 2013.[198]

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?

13. Provision of primary prophylaxis

Challenges in delivering primary prophylaxis

The barriers to delivering primary prophylaxis can be thought about in three main domains.

1. Patients and families

Sore throat is common, self-limited and usually a benign condition. Many people with a sore throat wait for symptoms to improve and do not seek medical care.

For example, in Tanzania parents of children with RHD did not routinely take children with a sore throat and fevers to seek medical treatment.[11] Similarly, qualitative interviews in at risk communities in New Zealand in 2013 revealed that families rarely sought assistance for sore throats.

Providing education and information about the importance of sore throat treatment is vital for effective delivery of primary prophylaxis.[11]

Even when people want to seek care there may be barriers which prevent access, including

- Cost of services or medication.
- Cost, difficulty and time of transport to health facility.
- Geographic distance.
- Household circumstances including weather, access to childcare and family commitments.
- Lengthy delays or excessive waiting times seeking care.
- Access to culturally appropriate health care.

★ Opportunities for research

Understanding the major barriers to care in your setting helps plan interventions- there's no sense making health clinics free if they're just too far away from where people live.

2. Individual health care providers

Health care providers must first know that GAS treatment guidelines exist. They must then be willing to adopt the guidelines. This knowledge and willingness may be influenced by training, education materials and professional experience with RF/RHD.

Even though using clinical guidelines seems to improve quality of care, clinicians are often unable, unwilling or unsupported to apply them in daily practice. For example, in Israel nearly 30,000 episodes of pharyngitis in almost 20,000 children were treated by 125 physicians from a range of specialties. Only half of the treating doctors followed national guidelines for treatment of pharyngitis.[233] In New Zealand only 80% of children with laboratory-confirmed GAS received appropriate management.[198]

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 guidelines recommendations into continuous quality improvement processes. Support clinicians to audit clinical practice against guideline recommendation.
- Dissemination involves making guidelines accessible, advertising their availability, and distributing them widely. Most evaluations of guidelines have shown that relying solely on printed material does not influence

clinicians' behaviour or health outcomes.[234] There is also evidence that unsolicited mailing of guidelines does not influence clinicians' behaviour, although it can increase awareness of the guidelines.[235]

3. The health system

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 systems' ability to respond to sore throats.[231] Practical health systems interventions can be helpful. For example, in New Zealand, a system to allow doctors to provide antibiotics for primary prophylaxis directly to patients (without have to visit a chemist or pharmacist) has been developed.[236] Similarly, children from high risk communities in New Zealand are offered school-based services for sore throat management. Children access trained lay workers sourced from the community and receive antibiotics from registered nurses, after parent consultation for allergies, for sore throat treatment.[135]

★ Opportunities for research

Understanding the major barriers to care in your setting helps plan interventions- there's no sense making health clinics free if they are just too far away from where people live to be accessible.

Box 11: The role of chemists and pharmacists

In many parts of the world private pharmacies are the main source of medication and health advice.[11] [237] 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. Fewer than 20% of the children had received medication from a dispensary, health centre or hospital.[186] In New Zealand a pilot project is underway to develop pharmacy-based sore throat management.[236] Including and educating private providers in your RHD control programme may be one approach to improve delivery of primary prophylaxis.

4.4. Active case finding: sore throat clinics

Background

randomised control trial encompassing 22,000 school
ildren was carried out in South Auckland, New Zealand
etween 1998 and 2001. South Auckland is an endemic
egion of disease with a large Māori and Pacific Island
opulation (RF incidence 60/100,000/year). Schools
ere randomised to receive a school based sore throat
inical or standard treatment. The intervention group
as diagnosed and treated for GAS pharyngitis by
urses in a school-based clinic programme. Treatment
ith a 10-day course of twice-daily oral penicillin,
dministered under nurse supervision, was initiated
ter a positive throat culture. Community health
orkers made daily classroom rounds to ask consented
ildren if they were experiencing symptoms of sore
roat, and performed monthly throat examinations
actively find asymptomatic cases. The control group
ceived standard general practice care outside of the
hool setting; management of the intervention group
d not include swabbing of symptomatic siblings who
ay have been in the control group. As GAS pharyngitis
well described as highly infectious this was a likely
ctor contributing to GAS pharyngitis load in the
mmunity.[239]

e endpoint measurement was incidence of acute
eumatic fever, determined by the Jones criteria,
borderline patients' not quite meeting criteria,
chocardiography was used to confirm or exclude the
iagnosis by screening for carditis. Results showed
a 20-30% relative risk reduction in ARF cases in the intervention group compared to the control group. However, these findings did not reach statistical significance and no clear recommendation could be made to advocate for the implementation of school-based GAS pharyngitis diagnosis and treatment programmes as a way to decrease incidence of RF.[240]

A subsequent meta-analysis that collated studies from New Zealand, Cuba, Hawaii, inner-city Baltimore and two American Indian reservations did, however, find a significant 60% reduction in incidence of RF conferred by school-based sore throat clinic programmes compared to general practice care. Recommendations based on this information were made to control RF in New Zealand as a developed country with significant RF burden in a disadvantaged population.[242] However, the conclusions of this meta-analysis have been challenged and application of results to other settings is unclear.[153] The cost and logistical challenges of introducing similar programmes in low resource settings may be prohibitive without considerable modification.[135, 161]

New Zealand Experience

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?

How would families and children consent for inclusion?

Would children who were absent or out of school in the programme?

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. Providing assessment and treatment of sore throats in a school based programme is an attempt to address these issues and maximise the impact of primary prevention.

The most comprehensive contemporary experience with sore throat clinics has occurred in New Zealand.[239] A small number of historic sore throat clinics were also run in the United States.[147, 240] Current evidence does not clearly demonstrate that this approach leads to significant reductions in the incidence of RF, but a broader programme is currently under evaluation in New Zealand.

Challenges in delivery of school based sore throat programmes

- An inconsistent number of children report sore throats preceding RF (see box 13).
- Understanding of GAS carriage is incomplete; children with GAS positive throat swabs will not necessarily have pharyngitis from GAS.
- Costs of delivering care may be very high.[132]
- Logistic challenges may be prohibitive in low and middle income country setting.[231]

Practicalities of school based sore throat programmes

The New Zealand Government has made a commitment to establish an extensive network of school sore throat clinics. In 2013 the programme included 211 schools, covering 47,500 children in high risk communities for school based throat swabs.[245, 246] New Zealand's sore throat programme is by far the most extensive internationally; therefore capacity to extrapolate considerations and recommendations to other settings is unclear. However, an overview of practicalities in the New Zealand model is presented here as a background for other programmes considering sore throat clinic development.

Consent

In the New Zealand model, families consent prospectively for throat swabs and treatment. Consent forms are signed by families in a two week window and all consented children are included in the sore throat programme.[247]

Throat swabs

In most New Zealand programmes Māori health workers visit schools a couple of times each week.[245] Younger children are asked daily during roll call whether they have a sore throat – older children are instructed to inform a teacher.[244] Health workers are supervised and supported by registered nurses.[110] The use of lay or community health workers has been an important component of historic sore throat clinics.[147, 240]

Results and treatment

Results from throat swabs are available within two days in New Zealand. The goal of the programme is for children to begin oral antibiotic treatment within 7 days of the onset of symptoms.[248,245] Children with positive GAS throat swabs are followed up by a registered nurse who can either take antibiotics to the family home or refer for a primary care consultation.[245, 247] Home visits provide an opportunity for health and hygiene education. In a previous United States programme children with positive GAS throat swabs were provided with a note to take home to parents advising them to seek medical attention.[240]

Integration

The New Zealand Government requires each District Health Board to have a senior executive 'RF Champion' to support integration within established health services.[245] Increasingly, comprehensive community consultation and strategic planning has increased capacity sore throat clinics to integrate with housing, skin sores and referral systems.[249]

Box 13: The challenge of low pharyngitis reports

The relationship between RF and preceding sore throats is complicated- a significant proportion of children present with RF without any recollection of pharyngitis. 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 17% had sought medical treatment.[97]
- In Pakistan only 30% - 40% of children had a strong history of sore throat preceding RF.[243]
- In a contemporary New Zealand setting, only 46% of children reported a sore throat prior to RF.[244] In another New Zealand study 14 of 19 children recalled a sore throat within 63 days of an episode of RF.[239]
- In Australia, 33% of all children with ARF reported a recent sore throat, reducing to 25% in Indigenous children.[60]

Poor correlation between sore throat and RF may represent **recall bias**, **asymptomatic infection**[16] or GAS infection from a skin source (see box 5, Chapter 5). If symptomatic GAS 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.

Things to consider

- existing intervention data
- How can you signal the need for a vaccine to governments or decision makers?
- Do you have enough information to advocate effectively?
- signal vaccine demand
- What other information will be needed to make decisions about vaccine delivery and implementation?

Development of an expensive vaccine is only worthwhile if there are no simpler or easier ways to achieve the same goal. Understanding what your country and programme spend on the existing interventions, and how well they work, is critical information for making vaccine investment decisions. Support from large stakeholders. RHD control programmes have the best possible insight into why a vaccine is needed and these experiences should be shared with governments, donors and decision makers whenever possible.

15. Vaccine development

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

Burchett et al, qualitative study of national-decision-making processes in seven low- and middle- income countries, 2012.[253]

A vaccine against GAS offers promise for definitive control of RF, RHD and other diseases caused by the same bacteria. Attempts to develop a GAS vaccine have been underway since the early 1920s and a number have progressed to early human trials.[95] Progress towards a safe, effective, affordable and practical GAS vaccine has accelerated in recent years.[250]

Prioritizing, developing and implementing vaccine programmes is a hugely complex international undertaking.[84] Similarly, basic science development of vaccines is highly specialised, expensive and technically complex.[250] These barriers can make it difficult for local disease control programmes to engage with global vaccine priority setting. In reality, 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.

The importance of early country level engagement is demonstrated by the rapid introduction of some vaccines (rotavirus and pneumococcal) after product licensure. Others, including HiB and Hepatitis B, experienced lengthy delays prior to widespread use.[251] In light of these experiences WHO developed Vaccine Introduction Guidelines to help countries make decisions about new vaccines.[252] This model has been expanded and adapted to explore preliminary work for a malaria vaccine, and a country planning for emerging health interventions.[253] A framework for preparatory GAS vaccine engagement is needed. In the interim key topics adapted from malaria are outlined in table 19.[253]

Burden of disease data	Burden of disease data is essential for demonstrating need and to inform decisions about potential benefits, cost effectiveness and impact. Locally measured data on burden of disease has a greater impact on decision makers than international estimates.[253, 254]

Case Study 5 | Nepal

Dr. P.R. Regmi | Cardiologist | President Nepal Heart Foundation

RF/RHD prevention in Nepal has been a joint effort between the government and the Nepal Heart Foundation (NHF) since 2008. Support and funding from the national government has made it possible to implement prevention policies on a wide scale throughout the country. The programme is run "diagonally" as a disease-specific intervention occurring within a developing health system. Dr. P.R. Regmi, executive director of the National RF and RHD Prevention and Control Programme and President of the Nepal Heart Foundation, shared his experiences:

Components of the programme:

Secondary prophylaxis is the major focus of the programme. Active screening of school children evaluates 10,000 students per year in three stages: 1) Clinical work-up including history and physical exam. 2) Patients exhibiting signs and symptoms consistent with RF/RHD then have their diagnosis confirmed or ruled-out via echo. 3) If confirmed, they are entered into a registry. Thirty-five participating hospitals throughout the country keep paper records of RF/RHD patients, which catalogue relevant clinical information including time of diagnosis, severity of disease and associated manifestations of disease, treatment given and reaction to penicillin. A central computerised database is kept in Kathmandu with compiled regional data. Patients receiving long-term secondary prophylaxis—in the form of BPG injections administered at district hospitals every 3 weeks—receive a "penicillin card" to document injection histories. The Nepal Heart Foundation procures BPG (and oral penicillin for patients unable to take BPG) and keeps a 6-month stockpile of the drug to protect against fluctuations in drug availability. Since the programme's inception, there has been no shortage of BPG.

The Lalitpur district in central Nepal is home to over 400,000 inhabitants with balanced rural/urban population distribution and was selected as a pilot site for a primary prophylaxis programme. Support comes from the government, the NHF, and Rotary International. Forty six health centres, health posts and sub-health posts in the region are involved and children ages 5-16 are targeted. Health professionals are trained to distinguish between viral and GAS pharyngitis based on clinical criteria; no swabbing or culturing is performed. Those children identified as having GAS pharyngitis are entered into a tonsilopharyngitis registry. Children are then treated with oral antibiotics. BPG is not viewed as a feasible treatment option due to resistance from both parents and physicians.

Linking sore throats to RF/RHD and seeking immediate treatment for sore throats are crucial components of the programme's awareness campaign. Audiences are targeted using multiple forms of media. Unique education initiatives include: a formal class for members of teachers unions, integration of RF/RHD into curricula through partnership with the Ministry of Education, a professional documentary that has proven very popular with the populace and has aired more than 50 times on national television, daily advertisements on morning radio, school children in Kathmandu performing street plays for the community, billboard, wall paintings and stickers.

A "Save Heart Everest Expedition" is being planned. Participants will hike from sea level in Bangladesh to the summit of Mt. Everest in approximately 45 days while relaying messages about cardiovascular disease and RHD.

Challenges and solutions:

Convincing the government of the need to target RF/RHD was difficult, in part because there are many other diseases and causes vying for governmental attention and assistance in Nepal. Presenting data in a simple, concise and understandable manner was important for influencing non-clinical decision-makers. Special attention was given to highlighting RF/RHD's disproportionate burden on the poorest sectors of Nepali society. Lobbying by the Nepal heart Network, a consortium of 19 heart NGO's, as well as doctors and local politicians was pivotal.

The fear of anaphylaxis, by both providers and parents, was a major hindrance at the outset. Providers experienced physical assault, jail punishment and demands for compensation after anaphylactic events suffered by their patients. These fears led many providers to halt BPG administration. In response, the NHF began training providers and developed protocols that outlined safe administration techniques. Emergency kits were provided and epinephrine syringes were to be loaded before each administration. NHF guaranteed providers legal support after an anaphylactic event if proper administration procedures were followed. Skin test protocols, which can actually precipitate an anaphylactic event, were stopped, except before first-time injections or if a change in brand or batch number occurred.

Moving forward:

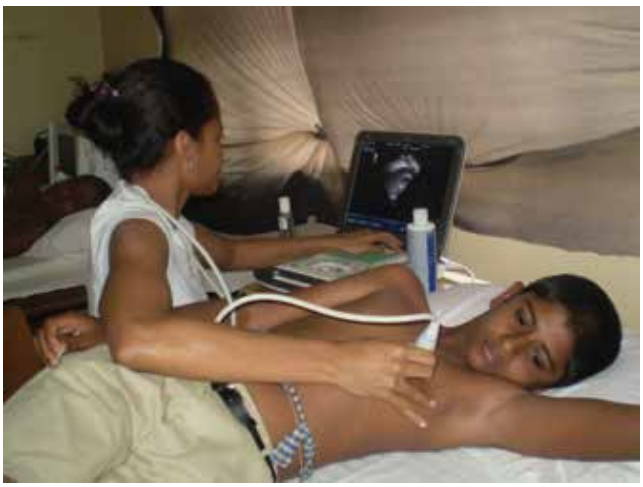
Although the programme is widespread in its distribution throughout the country, ideally the programme would be decentralised further to the sub-health post level. Currently, many individuals must travel to district hospitals to receive prophylactic penicillin every 3 weeks. The Ministry of Health must be convinced on a yearly basis to fund RF/RHD prevention. Preferably, programme funds would be guaranteed via its integration into a long-term non-communicable disease budget and run without the help from NHF and other outside organisations. One hundred percent of funds do come from the Nepal government, but even more funds are needed to implement certain programme objectives.

Secondary prevention has been an integral component of RHD control programmes since the development of effective antibiotics in the 1950s. Administration of the right antibiotics, at appropriate intervals, consistently over a number of years appears to prevent development of new GAS infections, and subsequent recurrences of RF. Preventing recurrences of RF slows, or perhaps even stops, the development of severe RHD. This disease-altering effect of antibiotic prophylaxis has been well described. The challenge is to support health systems to fulfill the promise of secondary prophylaxis.

Scientific understanding of the genetic, bacterial and environmental determinants of susceptibility to RF is imperfect. Only when young people present with clinical manifestations of RF is it possible to identify individuals at ongoing risk of RF, and subsequent RHD. This, necessarily retrospective identification of risk, creates a number of challenges. It requires accurate diagnosis of all cases of RF, and prompt enrolment into a register-based programme for antibiotic administration. Diagnosis and registration is a fragile process. Worldwide, many people are thought to be undiagnosed; because symptoms are mild, because families can not seek medical care, or because clinicians are unable to make an accurate diagnosis. Young people who are correctly identified as having RF then need to be enrolled into a register, to facilitate follow up, and ensure regular antibiotic administration. Even in places with an RHD register it may be difficult to enrol, families may not appreciate the importance of ongoing follow up or the register may be difficult for health staff to use. In other places, a register may not exist at all, making it all but impossible to deliver the years of antibiotic therapy which can halt disease progression.

In many parts of the world, secondary prevention defines the core business of RHD control programmes. Developing a register and delivering regular antibiotic injections has been a consistent feature of successful RHD control programmes over many decades. However, as outlined in chapters 1 – 10, secondary prophylaxis should not be delivered in isolation from the broader health system. RHD control programmes benefit from a comprehensive and integrated approach. In some cases, integration makes practical sense (identifying cases through notifications, collecting epidemiologic data to understand the burden of disease), in other domains it can foster sustainability (developing advocates and strengthening fundraising). Integration with primary and tertiary interventions can similarly amplify the role of registers by reducing the burden of new cases, and improving delivery of care to people already living with RHD. Register based secondary prophylaxis appears to be a necessary precondition for RHD control, but it is unlikely to be sufficient as an isolated intervention.

A register and schedule for secondary prophylaxis should be one of the first priorities for new and redeveloped RHD control programmes. These services should be delivered to a demonstrably high standard before more complex interventions – for example, surgical programmes, echocardiography screening for clinical benefit –begin. If the goal of active case finding is to change clinical outcomes, it is reasonable to ensure that disease altering interventions can indeed be delivered. Similarly, the vast majority of national guidelines recommend sustained secondary prophylaxis after operative intervention. Health systems should be expected to deliver this baseline care pre-operatively, before post-operative planning can meaningfully begin.



Echocardiography in Fiji

Things to consider

Do you have an RF/RHD register?

Where is it kept and how is it maintained?

How are people added to or removed from the register?

How will their information be kept confidential?

How is register data communicated to public health or planning groups?

Is the register used for research purposes?

16. The RF and 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.[20]

RF/RHD registers - a list of people with RF/RHD and some of their clinical details- were established in the United States in the 1950s.[62] These registers helped provide newly developed regimes for antibiotic secondary prophylaxis and contributed to the declining burden of RHD in the USA.[62] By the 1970s the WHO had adopted a register based approach and registers remain a critical part of RHD control.[223 231, 255] Register based programmes have assisted with the provision of prophylaxis in many communities across the globe, including New Zealand,[256] Australia,[16, 158, 257] Samoa,[72] Fiji,[257] Cuba,[26] Egypt.[258]

Why is a register so important?

An RF/RHD register can assist with routine assessment and surveillance. It is useful for recording prophylaxis delivery, the recall of patients who are due for, or miss doses of, BPG, and informing health education and health promotion programmes. Registers also provide some information about the burden of disease; though the quality of data is dependent on the quality of register management.[29]

Box 14: Register based programme example Northern Territory, Australia[187]

The Australian Northern Territory (NT) RHD Control Programme has a central register with a master list of patients. Data relating to the provision of secondary prophylaxis in primary health centres are stored locally within local registers and within case notes.

Primary health workers mail, fax or email a form monthly to the central register with details of who has received secondary prophylaxis and who has missed a dose. Data are entered into the central register by RHD programme staff and epidemiological reports are produced for the Programme Advisory Committee and sent to each of the primary health centres for the review of local staff.

The NT control programme received funding in 2007 to develop an internet based register that enables local primary health workers to log on to a secure website and enter and view information relevant to their location. Due to the challenges of internet access in remote communities and time constraints some still find it convenient to send the information monthly to the control programme staff for input.

Improves delivery of consistent, disease altering, secondary prophylaxis through:

- Recall systems
- Standardised care delivery
- Helps to identify people with poor adherence for additional support
- Provides information about the burden of disease over time
- Facilitates monitoring of recurrence rates and other indicators

Electronic

Paper

	Advantages	Disadvantages
Register logistics	• Reports and data easily extracted • Can be updated and accessed by local clinical staff	• May be more expensive to establish and maintain • Can be difficult to transmit information to a central register, unless a computer is available • Data may be lost
Location	• Little training required • Valuable for mobile populations or people who move frequently	• Requires systems to provide information back to the health system level • May not be clinically relevant for primary care provision

Two major decisions are needed about your register: location and format. Advantages and disadvantages of options are summarised in tables 21 and 22.

Location

Central registers

Centralised registers can support the provision of prophylaxis for those who move between communities and provide valuable epidemiological data to assist public health clinicians, managers and bureaucrats to understand the disease burden in different areas. This assists with the allocation of resources to where they are needed the most. Only some of the information stored on the local register may need to be provided to the central register. For example, in Nepal local registers record people admitted with RF/RHD and summary lists are forwarded to the central (national) register maintained by the Nepal Heart Foundation.[258]

Local registers

A local register can be a paper based list of people living with RHD in the community. The person will need to be followed up (reviewed and treated) on a regular basis, possibly for many years, therefore the register is a way of keeping track of what is required and when. It may need to be reviewed daily or weekly to ascertain who needs to be recalled and who is overdue.

There may need to be several components, potentially including: next secondary prophylaxis injection date, clinical review schedule and dental follow up. As your programme develops these can be formalised into priority based follow up and personalised care plans (see Chapter 19).

In practice, most settings have a local register to record adherence and send information to a central state/country register where trends are monitored over time.[256]

Register format

Paper registers

Details recorded on paper cards and stored in a container with tabs for each month of the year or filed in another way that suits the local situation. The cards can be placed in the month that the person requires their next appointment and their clinical file accessed for further information.

One of the challenges of paper registers is ensuring that data are protected from loss or damage. In Samoa a misplaced paper register reduced the effectiveness of the RHD control programme.[259]

Computer registers

A computer register may be a unique data base utilised only for RF/RHD control and management, or one that is part of a patient information system that is used for all patient management. The important aspect of a computer register is that it has the required fields to store information within it and 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, and that it can be easily used by staff.

Although computer registers are more durable than paper copies (and more easily backed up) software and hardware maintenance is required. Changes in file types and software platforms can make it difficult to extract retrospective data.[95]

Establishing a register

Minimum data set

There is no current global consensus about what information should be collected and stored on an RHD register. Some programmes have established their own data standards.[28, 261]

Privacy, confidentiality and data security

Use, ownership and protection of health data are complicated issues worldwide.[62] Many settings have struggled to establish protocols to manage confidential health information. For RHD registers, privacy requirements may influence which data is collected and how it can be used. It is helpful to seek advice as early as possible when establishing or running an RHD register to ensure that your programme complies with local laws, standards and procedures. You may need to seek input from ethics committees, health authorities or other register based programmes. Data which is accidently collected inappropriately (without sufficient consent or safeguards) may not be able to be used.

Education and training

Staff will require training to use and maintain the RHD register. The more complicated the system the more time it will take for people to become familiar with it. Integrating the register within existing systems will limit the number of systems new staff will need to become familiar with.[20]

★ Opportunities for primordial prevention

Some programmes choose to include factors relevant to primordial prevention on registers, including socioeconomic indicators, children in the family or other details.[262] Even when it is not possible to demonstrate causation, understanding the demographic and socioeconomic characteristics of people presenting with RF/RHD is helpful for targeting services and intervention for primordial prevention.

Maintaining 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."

Fiesa et al, Northern Territory, Australia, 2005.[66]

The RF/RHD register is only as good as the information entered. Complete, high quality data is critical for programmatic success. Your programme will need a clear protocol for ensuring that people can be added to the register. There will likely need to be multiple entry and change points within the register, a sample of these are outlined in table 23.

After an episode of RF	Prophylaxis ceased or completed
People at risk of RF/RHD should be enrolled 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, that the register exists, contact details and the information required for enrollment. The date of first symptoms (rather than the date of clinical presentation) should be recorded. If possible it is simplest to encourage clinicians to contact the register about all cases of RF (suspected and confirmed, first episode and recurrences). This reduces the decision making burden for individual clinicians and provides as much information as possible to your programme. Information about recurrences 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.[263]	Treatment and management guidelines should be clear about the duration of secondary prophylaxis. The person's history of RF and the presence of heart valve damage associated with RHD will guide the decision to cease secondary prophylaxis. Injections of BPG - or other prophylaxis- should be stopped when clinically appropriate. Systems are needed to make sure that prophylaxis is reviewed when treatment is complete. The decision to stop treatment by specialist clinicians needs to be clearly communicated to teams responsible for administering the 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.[66] The register should record whether the planned duration of secondary prophylaxis was delivered (according to local guidelines), whether treatment was stopped early following expert clinical review or whether the treatment was stopped without clinical consultation.
Clinicians who diagnose RHD need to be able to contact the register coordinator to enroll people living with RHD into the register. Advanced cases of RHD may be identified late in adults and, rarely, the elderly. Therefore, adult clinicians, midwives, and primary care staff will need to know to contact your control programme. Ideally, clinicians will also 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.	All programmes will lose some people to follow up – due to unreported deaths, unplanned travel, and unplanned changes in contact details or active avoidance. These people continue to be epidemiologically relevant, 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 for data to be retained without active care delivery. You will need to define 'lost to follow up'.
Echocardiographic screening programmes (outlined in Chapter 19)should be closely aligned to the RF/RHD register to ensure that people with RHD identified during screening receive appropriate care and follow up.	Recording deaths of individuals on the RHD register is important in order to: • Avoid distress for families and communities by following up deceased individuals inappropriately. • Ensure that resources are not consumed attempting to follow up individuals who are no longer alive. • Understand the mortality burden of RHD and develop a sense of local disease prognosis, particularly if information about cause of death is available. • Compile a record of surgical outcome audits. In some places it may be possible to access hospital death records to identify people with RHD who have died.[158] 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. As much information as possible about the cause of death should be recorded.
In some countries people living with RHD are a mobile population who move frequently for work, health care, family or traditional responsibilities.[20,23,66] You will need a protocol for accepting registrations from other programmes and for enrolling 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.[60] Other sources of information for the register may include: notifications, hospital discharge records, clinical letters, echocardiogram reports or professional correspondence.[66, 158] These records may also provide valuable information about the clinical status of people already on the register.	People on your register may need to move outside the scope of your programme. When travel or relocation is planned or expected you should develop plans to 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 travel. • 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 World Heart Federation 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'.[5] Although the threshold criteria disease elimination and control at national and regional levels are still being developed 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; identifying criteria for closing registers long in advance may help prevent premature programme closure.

The best information about closing/phasing out register based programmes comes from the United States in the 1970s.[264] In 1977, 29 states of the United States had some form of RHD register; by 1979 only 11 of these states had ongoing register based programmes. Reduction in programme numbers was attributed to an apparent decrease of cases of RF and budget constraints.

Retaining some mechanism for monitoring of RF/RHD is advisable, even when disease control targets have been met.[265] A background rate of disease persists in high resource settings and disease resurgence after a period of control is possible.[266] For example, in Utah an eightfold increase in cases of RF occurred in 1985.[97] Although this cluster of cases had some atypical features, the outbreak demonstrates the importance of maintaining some mechanism for identifying and managing RF and RHD.[97]

Box 15: Expect an apparent increase in cases when notifications and registrations begin

"The recent discontinuation of the Waikato register shows that previous

Starting or strengthening your control programme will tend to increase health worker and community awareness about RF. This may make people more vigilant, prompting notification of suspected cases of RF which may otherwise have been overlooked.[22, 30] A surge of interest can sometimes make it appear as though there is a new epidemic of disease. For example, in the French Caribbean: "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".[26]

Case Study 6 | Egypt

Dr. Alaa Elghamrawy | Consultant Cardiology | Director, RF/RHD Prevention and Control Programme

"The Rheumatic Fever/Rheumatic Heart Disease Prevention and Control Programme in Egypt was started in 2002. Partial financial and technical support was provided by WHO, but the programme has been facilitated by the Egyptian Ministry of Health. Programme Director, Dr. Elghamrawy, spoke about his experiences:

Components of the programme

RHD is attacked from a variety of angles with a focus on both primary and secondary prevention. In addition, because the programme is run via the Ministry of Health, an emphasis is placed on capacity building, both in the public health and medical sectors. Hundreds of health workers are trained to recognise the clinical signs and symptoms of GAS pharyngitis and to administer proper treatment with one-time BPG injection. Training materials, including clinical guidelines, are distributed to health professionals down to the health unit level where providers are instructed to refer patients with symptoms for formal evaluation with echo at a regional center.

Case finding is an active component of the programme. Screening campaigns with portable echo have yielded thousands of cases of RHD. Surveys of school children for signs and symptoms are also used to detect cases. Medical caravans are used to reach children in remote areas. Those identified as having RHD are entered onto a registry. Five regional centers have their own electronic databases. Data from these five centres are pooled at the Ministry of Health into a single, central registry. To date, over 8,000 patients are registered.

Awareness campaigns target a range of populations from health workers to the public at large. Religious leaders, youth centre staff and teachers are specifically targeted in community outreach campaigns. Brochures are created for widespread public dissemination. Media outlets, including radio and television, are also used for mass education of the public.

Challenges

The budget is insufficient and requires individual fundraising efforts. Financial support from WHO comes only every two years, and the Ministry of Health contributes mostly by providing human capital rather than monetary assistance. A stand-alone budget for RF/RHD prevention is needed in order to reduce dependence on funding from outside sources. Initially, there was a disconnect between government and NGO stakeholders. It is believed that partnerships could be better coordinated to maximise impact.

With current funding levels, it is difficult to maintain a consistent level of services. Increasing the scaling of the project has also been difficult. At the programme's inception there were two referral centres. Although five referral centres exist today, it is still an inadequate number to sufficiently address the disease burden. Hardware shortages at these facilities restrict capacity, and software for maintaining a register is outdated. Additionally, echo machines require servicing and maintenance

Continuity of medical care is hampered by the high turnover of primary care staff. Frequent transfers and departures necessitate the continual training of health care professionals. Regular support from WHO is required to ensure this training. In addition, maintaining physician motivation is difficult without financial incentive.

Moving forward

The Egyptian experience is being exported to some neighbouring Arabic countries. Disease burden attributable to RF/RHD is very high throughout this region.

Acknowledgement: Dr. Alaa Elghamrawy would like to express appreciation for the help of Dr. Mokhtar Gomaa, Professor of Cardiology (Al Azhar University) and Dr. Fathy Meklady, Professor of Cardiology (Suez Canal University) in the successful completion of the programme.

Is BPG on your national essential medicines list or formulary?

Do you have stock outs or shortages of BPG?

Do you ...e?

Do you ...m for ensuring all people are asked about a history of penicillin allergy?

Do you ...ing for management of anaphylaxis?

Do you ...to adrenaline and other treatments for anaphylaxis?

Do you ...anisms for monitoring or reporting adverse drug reactions?

Table 24: Formulations of BPG

Liquid	Currently produced under patent by a single manufacturer, dispensed in a prefilled syringe and relatively expensive. The liquid formulation requires refrigeration and is most widely used in high income settings.
Powdered	Powdered forms of BPG are produced by a number of different generic manufacturers and are relatively inexpensive. The sterile powder must be mixed with sterile water prior to injection. It does not require a cold chain and can be stored for a number of years.

BPG and other antibiotic supply

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

Benzathine Penicillin G

The antibiotic benzathine penicillin G (BPG), also known as benzathine benzyl penicillin, is commonly used for primary and secondary prophylaxis. BPG was developed in the 1950s as a relatively insoluble penicillin which is injected intramuscularly.[267] Low solubility of BPG means that penicillin remains in the blood for weeks, preventing GAS infections. Globally there are two existing formulations of BPG, outlined in table 24.[267-269]

Supply of BPG

Stock outs and shortages of BPG have occurred in the United States, Canada, Australia and New Zealand in recent decades.[268, 270-273] Consistent anecdotal reports from low and middle income countries were investigated by the World Heart Federation in 2011.[274] From a survey of 39 respondents almost all reported, 'minimal' access to BPG and 35% reported inadequate supply to treat patients according to a recommended schedule of secondary prophylaxis injections.[274]

Responding to stockouts

In the event of a shortage or stock out of BPG the World Heart Federation recommends:[20]

- Confirm when BPG will be available, and emphasize to the responsible authorities the critical importance of ensuring supply as soon as possible.
- Redirect existing supplies to health facilities with the highest demand.
- Communicate with health facility staff and recommend that oral penicillin be used until BPG is readily available.
- Health staff should clearly inform people who usually receive BPG that oral penicillin is only a temporary solution, and that they will be recalled when the BPG injections become available.

Quality of BPG

There are two main quality concerns about powdered, generic formulations of BPG

1. *Difficulty mixing the powdered BPG into a suspended solution.*
   Persistent anecdotal reports suggest that generic powdered formulations of BPG are very difficult to suspend in solution.[266] This causes blockages in the needle during delivery, apparently increases pain and may result in inaccurate dosing.

2. *Duration of serum penicillin concentration levels.*
   There is some research to suggest that the serum concentration of penicillin falls more quickly than expected.[267, 275] The cause of this declining serum concentration is unclear and complicated by difficulties in identifying and monitoring the generic manufacturers of BPG. There are no readily available manufacturing standards or assays for analysis of powdered formulations.[267]

Safety and anaphylaxis

Patients and health care providers are understandably concerned about the risk of anaphylaxis from BPG injections.[34,150, 168, 266, 275, 276]

The best information about adverse reaction and allergy to BPG comes from a paper by the International Rheumatic Fever Study Group in 1991. Between 1988 and 1990 they considered 1790 people from 11 countries having 32,340 injections of BPG. [277]

In this study:

- 57/1790 people (3.2%) had an allergic reaction
- 4/1790 people (0.22%) had anaphylaxis (= 1.2/10,000 injections)
- 1/1790 people (0.05%) died (= 0.31/10,000 injections). This single death occurred in a 15 year old patient with severe mitral valve disease, and congestive heart failure.

Overall, the disease altering benefit of BPG injections outweighs the small risk of a fatal allergic reaction.[21, 278]

Defining anaphylaxis

'Anaphylaxis is a severe, life-threatening, generalized or systemic hypersensitivity reaction.[278, 279] The international diagnostic criteria for anaphylaxis are outlined in Annex C. Although there has been considerable concern about anaphylaxis from BPG it remains unclear whether adverse events have been genuine hypersensitivity reactions. Some reported cases do not have a lot of features of anaphylaxis (e.g. swollen airways, difficulty breathing) and other causes (such as contaminated batches of BPG, inadvertent IV injection, or vasovagal episodes in people with severe pre-existing valvular disease) have not always been ruled out. Therefore, the issue of anaphylaxis should be thoroughly investigated before embarking on extreme measures such as routine skin testing[45, 46] or even banning BPG.[276]

Management of anaphylaxis

Managing the perception of risk from BPG injections is an important part of education and support for an RHD control programme. Deaths thought to be associated with penicillin can dramatically undermine community engagement, and risk more lives if people living with RHD cease prophylaxis.

Strong protocols for administering penicillin and managing adverse drug reaction are needed to deliver the medication safely. Wherever possible, 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.[279] Training and equipment will be needed for effective management of anaphylaxis.[26, 63]

★ Opportunities for integration

More than 65 countries have some kind of medicines agency or **pharmacovigilance** programme.[280] 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 World Health Organization and partners.

Box 16. Local anaesthetic for IM injection pain in BPG prophylaxis

Some programmes mix BPG with local anaesthetic to try and reduce the pain of administration.[643 284, 285] There is reasonable evidence that using local anaesthetic reduces pain without compromising serum concentration of BPG.[286-288] However, this practice is not currently licensed, nor supported by BPG manufacturers.

Although there is conflicting evidence about how much pain from injections impacts on patient adherence, it is reasonable to seek to minimise discomfort.[282] A number of programmes have developed injection protocols or guidelines to reduce pain.[20, 29, 63, 283]

A summary of strategies:

- Use a 21-gauge needle – smaller needles are much more likely to block and increase pain during administration.
- Warm syringe to room temperature immediately before using.
- 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.
- Apply pressure with thumb for 10 seconds before inserting needle.
- Deliver injection very slowly, preferably over at least 2–3 minutes.
- Distract patient during injection, for example, with conversation.

Securing supplies of BPG

Defining BPG as an essential medicine

BPG is on the World Health Organization International Essential Medicines List, the Essential Medicines List for Children and the Essential Medicines List for Sexual and Reproductive Health.[288, 289] However 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 EML/formulary. Some low and middle income countries are eligible to purchase medication from UNICEF or the International Dispensary Association. Information about reasonable prices for BPG (and other medications) can be obtained from the International Drug Price Indicator Guide, produced by Management Sciences for Health each year.

Making BPG affordable for people living with RHD

Financial cost may be 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.[290] A number of successful RHD programmes have secured a supply of BPG which is free to some or to all patients including programmes in the French Caribbean, the United States, Nepal and Pakistan.[27, 63, 264 292, 293]

Other antibiotics

A small minority of 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.[294] See summary, Chapter 20.

★ Opportunities for integration

BPG is also used in the treatment of yaws and some forms of syphilis (although sometimes in other doses). You may be able to work with yaws or syphilis teams to improve the supply of BPG.

★ Opportunities for research

The real cost to consumers of receiving secondary prophylaxis is sometimes unclear. An audit of pharmacies and health centers may provide valuable information about the kinds of BPG being sold on the market, and variations in price. Supply of BPG is a worldwide challenge, and there is insufficient information about fluctuations in price and availability. Could your programme document prices and stockouts (or encourage people receiving secondary prophylaxis to maintain their own records) in order to better understand the problem?

"Undoubtedly, the main reasons of the severity of disease in Yemen are: the absence of a specific program for the prevention and control of RHD, the lack of government decision to adopt such a program and the inadequate use of penicillin by general practitioners because of fear over allergic reactions or lack of precise information regarding it's indication, dose and duration."

Saleh, Yemen, 2007.[277]

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 you have an automated recall system for people overdue for secondary prophylaxis injections?

18. Provision of secondary prophylaxis

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. The two international guidelines come from WHO[21] and WHF.[120]

Local development and guideline adaption has occurred in Australia,[28] New Zealand,[260] Western Cape South Africa[294] and India.[295] You will need to adopt and adapt one of these guidelines for your country or programme.

A summary of secondary prophylaxis guidelines appears in table 25.

Table 25: Summarised secondary prophylaxis guidlines

Guidelines	Preferred antibiotic	IM BPG doses	Interval of BPG injections	Oral alternatives	Duration of therapy	Year
WHO[21]	BPG	<30kg: 0.6U >30kg: 1.2U	21 days if high risk 28 days if low risk	Pen V** 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[29]	BPG	<20kg: 0.6U >20kg: 1.2U	4 weeks (3 weeks for selected groups)	Pen V** 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
India[296]	BPG	<27kg: 0.6U >27kg: 1.2U	<27kg: 15 days >27kg: 21 days	Pen V** Children:250mg 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[295]	BPG	<30kg 0.6-0.9U >30kg: 1.2U	3 weekly	Pen V** <30kg: 125mg twice daily >30kg 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. Severe/post valve surgery: lifelong	1997

* Whichever is longer

** Pen V = Phenoxymethyl-penicillin

Box 17: Defining "non-adherence"

esearch into determinants of secondary prophylaxis
dherence requires a common measure which is
omparable across countries. Using a common metric
akes it easier to establish determinants of adherence
nd to generalize these results to other settings.

roposed measures of adherence are:

The proportion of scheduled BPG injections delivered in the previous 12 months

Individual and community figures expressed as:

Median percentage of doses delivered

Proportion who receive 80% or less of scheduled doses

Proportion who receive 50% or less scheduled doses

hese measures are really indictors of service delivery
nd quality. They should not be confused with a biologic
easure of protection from GAS infection, nor should an
0% threshold be considered 'good enough'.

00% adherence with scheduled injections is the most
ffective approach to secondary prophylaxis.[299,300]

Supporting adherence

Adherence to the local schedule of secondary prophylaxis injection is poor worldwide.[295] Adherence is consistently less than 50% of scheduled injections delivered in a range of settings including Indigenous Australia, Brazil, Ethiopia, New Caledonia, South Africa and the Congo.[68, 81, 238, 297, 262, 298]

New Zealand is one setting with sustained, high quality delivery of secondary prophylaxis. The Auckland Rheumatic Fever Register was established in 1981 and has been broadly supported by community health workers and trained nursing staff.[192] By the year 2000, all local programmes reported full adherence greater than 86%, rising to 96% in some areas.[192] Similarly, in Samoa, adherence with BPG injections was less than 50% between 2001 - 2006. Adherence rates improved to 74% and 84% after government intervention and a re-invigorated RHD programme.[192]

Box 18: Language matters

Internationally, the term 'compliance' is often used to describe whether or not individuals who are 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.

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

McDonald, Australia, 2004. [302]

Using language which reflects shared responsibility for health may help reduce stigma and frustration with individuals perceived to be 'non-compliant'. In this handbook we have used the term 'adherence' to refer to delivery of scheduled medication.

Contributors to poor adherence

There are many reasons for individuals to miss scheduled BPG injections. Key determinants are outlined below and a systematic evaluation of interventions is underway in Australia.[302]

Perception 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.[282] 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.[298] Education may be particularly important for people incidentally diagnosed with RHD (see Chapter 11, Community Education). Cultural beliefs, including the role of traditional medicines (see box 25, page 79) may also contribute to adherence behaviours.

Experience of care delivery

The experiences that people have when receiving injections (and in the broader health system) determine whether they will keep returning for injections. Qualitative studies suggest that supportive relationships with clinical staff encourage trust and support return attendance.[63,66]

Cost

In some places there is a direct financial cost to receive BPG injections (See Chapter 17). In other places people must take time off work or school to travel for injections causing indirect costs in time and travel. Cost and travel have been identified as barriers to adherence in Ethiopia,[199] India,[282] Thailand,[304] and Egypt.[291] In Thailand in the 1980s approximately 200 people receiving monthly BPG injections were randomised to centralised dispensing (main hospital pediatric department) or decentralised dispensing (local health centres with 3 monthly specialist 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.[304]

★ Research opportunities

Understanding the determinants of poor adherence in your setting can be helpful for improving service delivery. You may be able to identify people who have been lost to follow up and explore the factors associated with poor adherence.

Strategies to improve adherence

Employ or identify regular staff to deliver secondary prophylaxis

A single dedicated health care worker responsible for administering BPG and ensuring adherence seems to improve uptake.[67, 158, 191] Building relationships with trusted health care professionals appears to be an important determinant of adherence.

Produce prophylaxis cards

A card recording BPG administration has been a popular option for documenting administered injections and recording the date of the next appointment.[39, 238, 305, 306] In the early years of the WHO control programmeme BPG cards were produced and countries were encouraged to adapt their own format.[100] The contemporary Pacific RHD Control Programme also uses personal medication cards, adapted to local language and needs.[162] Communities in some settings are very familiar with personally held medical records. In other countries cards are used for monitoring health delivery and outcomes, including immunisation cards and 'under 5' cards.[307, 308]

Support 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.[192, 238, 309] In Perth (Australia) and Auckland (New Zealand) this approach has been associated with high rates of adherence.[192, 309] Programmemes considering mobile injection delivery should consider the safety of health care workers (including dogs, violence and travel) as well as their capacity to manage adverse drug reactions in the community.[309]

★ Opportunities for primordial prevention

Home visits provide an opportunity to evaluate living conditions, and provide education to family groups.

Memory cues

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.[310] 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 a more consistent uptake during the full moon.[310] Other opportunities may include provision of calendars or the development of a smartphone app.

Text messaging and phone calls

Text messages (SMS or texting) can be used to remind people that injections are due.[72, 311] There is some evidence that text messages as health reminders can improve appointment attendance.[312] The effect on secondary prophylaxis delivery requires further research but offers

"Due to better access to hospitals and improved transportation infrastructure around the island, it is now considerably easier to get the IM penicillin injections. The key to better compliance was enthusiastic, dedicated staff in the Rheumatic Fever Programme and the regular patient call to remind the patients of the injections."

Viali et al, Samoa, 2011.[72]

conceptual promise. In the Pacific Islands phone calls or long distance radio messages have been used to encourage people to return for secondary prophylaxis delivery.[313]

As with other messages and social media campaigns, text messaging should be part of a larger integrated communication strategy. The Centers for Disease Control suggests considering:[313]

- The availability and use of mobile/cellular phones in the target population.
- Length of characters available for use in the message (160 characters using the English format and less in many other languages- Arabic, Chinese, Korean, Japanese, or Cyrillic alphabet languages.[315])
- Develop a protocol for refining the messages to ensure that the message impacts the target.
- Be clear about the objective of the campaign, including the overall objective, and the action the campaign is wanting the recipient to take.
- Budget for sustainability.
- Integration of the campaign requires communication with other agencies that will be impacted by the action taken by the recipient – be sure other agencies are prepared for the actions.

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 centers. This may be "convenient as far as time and travel expense are concerned".[292, 293] 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.[43] 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.[236, 316]

Developing a recall system for missing patients

Early recall systems may be helpful for reminding patients to have each injection, and to identify people who have been lost to follow up before they have moved too far away. In the Northern Territory of Australia the recall notifications have recently been moved from day 28 to day 21 after each injection to attempt to improve adherence.[317]

Box 19: Supporting adherence with secondary prophylaxis is everyone's responsibility

"Every contact with a health professional that does not discuss secondary prophylaxis is a substandard consultation"

Wilson, New Zealand, 2013.[299]

Consistent messages about the value of secondary prophylaxis should be provided to all health care staff, communities and people living with RHD. All staff should be empowered to discuss adherence. 100% of scheduled injections should be the goal for every person receiving secondary prophylaxis.

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?

19. Priority based follow up (clinical review)

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. The register will include people living with RHD at very different stages of disease - some will have no symptoms, others will have severe disease or end stage heart failure. The clinical needs of these patients are different and a system is needed to ensure that patients who need the most input receive the closest follow up.

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.[20, 31, 49, 62] Follow up guidelines need to be developed by each programme to reflect local resources, distribution of services and opportunities to connect with surgical services where required.

- 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.
- Data can be utilised for epidemiological 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 clinicians 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. Establish 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 **NYHA** classification and the Australian 'priority based' follow up categories.

2. Establish follow up recommendations

In settings where a primary health system is established, follow up recommendations should be undertaken in consultation with family doctors or health centres. Ideally, follow up activities are integrated into the activities of primary health workers, with 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 on evidence that is applicable to the setting.

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. Plans should be integrated and recorded within the patient information and recall system, or local health care record.

A recommended and routine review and management plan (a care plan) can assist clinicians with assigning a management plan to patients based on the level of disease.[29, 318] Providing a standardised case management approach has been valuable for other conditions endemic in low resource settings.[109, 319]

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 and 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 or vaccination.

Implementing priority based follow up

Strengthen referral systems

Referrals between primary, secondary tertiary and quaternary health care providers are a risky period for loss of clinical information or breakdown in continuity of care.[30, 67] 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.[199, 305] Transport may be difficult or prohibitively expensive.[199, 305] 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 the 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, United States Associated Pacific Islands, 2008.[313]

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?

20. Active case finding (echocardiography screening)

An introduction to screening

Health screening programmes are designed to 'discover those among the apparently well who are in fact suffering from the disease'.[320] 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.[65] A WHO supported auscultation screening programme began in 1984, and included 1.4 million school children in 16 countries.[321] In 2001 the WHO Expert Committee for RF and RHD again recommended auscultation screening for high risk populations.[21] This search for heart murmurs represented a large scale attempt to identify children with RHD. However, the advent of echocardiography (echo) has revolutionised screening for RHD, offering risks and benefits.[322]

The role of echocardiography in screening for RHD began to be explored in the mid-1990s.[181, 322] 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 echo screening compared with auscultation screening.[45] Since then a large number of echo screening projects have been undertaken around the world, including in Tonga,[323] India,[324] Nepal,[284] Nicaragua,[325] New Caledonia,[56] New Zealand[180] and Uganda.[327]

Most large scale echo projects have been conducted to provide baseline descriptive epidemiology and burden of disease data. The information and engagement from echo screening projects has been important for the growing international interest in RHD.[328] However, the clinical application of echo screening for detecting early disease and providing opportunities for intervention remains unclear. The rapid increase in echo screening programmes has prompted concern about the ethics and feasibility of screening, given these uncertainties.[322, 329]

Many countries have criteria to establish when population screening is appropriate, and what issues need to be considered. Sample critiera are presented in table 27.[320] The appropriateness of echocardiography screening for RHD has been addressed by a number of authors and is summarised in the table 27.[47, 322, 329]

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

Remenyi et al, The World Heart Federation Criteria for Echocardiographic Diagnosis of RHD, 2013.[45]

Although echocardiography is a relatively non-invasive procedure, [illegible] disease has been challenging. There are many small variations of normal heart valves, and it may be difficult to distinguish normal heart valve function from rheumatic heart valve damage. Early studies of echo screening for RHD used slightly different criteria to define disease, making it difficult to interpret and compare results from around the world.[46] Despite attempts to standardise diagnostic criteria by WHO, large variations in burden of disease were caused by subtle changes in criteria for diagnosis.[322] In 2012, a group of experts developed a rigorous approach to diagnose subclinical RHD: the World Heart Federation criteria for echocardiographic diagnosis of RHD.[45] These criteria define the morphology (shape) and functional changes of valves affected by RHD.

The natural history of subclinical RHD has not been fully established, and the progression, stability or regression of valve lesions is unclear.

Box 20: The role of auscultation screening

Where echo is available, screening for RHD by auscultation alone is not appropriate.[322] A study of 1015 high risk Indigenous school children in the Northern Territory of Australia revealed unacceptably poor sensitivity and positive predictive value.[331]

Table 27: Suitability of RHD for echocardiography screening[320]

Evidence of a significant burden of disease	There is strong evidence of a significant burden of disease in low resource settings worldwide.
Condition must have a latent stage	RHD has an asymptomatic phase, particularly if the first episode of RF was mild or undiagnosed.[4] Mild valve lesions identifiable only through echocardiography are called 'subclinical RHD'. The clinical significance and natural history of valve lesions in RHD are poorly understood.
The early stage of disease must be treatable with adequate therapy	Regular secondary prophylaxis prevents recurrences of RF, and minimizes progression in valve lesions of RHD. However, delivery of high quality prophylaxis is difficult in many parts of the world. If your programme is unable to provide very good secondary prophylaxis to people already on the register, undertaking ongoing clinical echo screening for RHD is rarely appropriate or ethical.

Echocardiographic diagnostic criteria for RHD

The World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease are the universal diagnostic standard for defining of RHD in people less than 20 years of age.[45] For screening, a simplified criteria that can be utilised by technicians with basic levels of training in echo may be used prior to expert radiologic and clinical review.[330] All screening programmes should include expert review of images according to the World Heart Federation criteria.

early detection of RHD in childhood that includes non-school goers will have to be developed”.

Maharaj and Parrish, Prevention of infective endocarditis in developing countries, 2012.[138]

Who should be screened?

Age

The age of people being screened has a significant impact on results. In general, as the screened population is older (usually late adolescence) more cases of RHD will be identified and valve lesions tend to be more severe.[47, 166,332] These more advanced cases of RHD may offer relatively little opportunity to intervene and prevent disease progression.

School children

Schools can provide a convenient way of accessing large numbers of young people in a single location. However, school attendance varies around the world; in some places children at greatest risk of RHD may be the least likely to attend school, or be accessible for screening.[38, 138]

Models of screening

The emerging protocol for echo screening studies is a short, targeted echo of the mitral and aortic valves to assess the WHF criteria for RHD. People with a suspicious screening echo need to have a second, more detailed echo, to provide a final decision on diagnosis.

Who should do the screening?

The training and skills required to undertake appropriate echocardiography screening are the subject of ongoing investigation. Some studies have used medical students[120, 333] or doctors without cardiology training as informal echocardiographers. A formal pilot study to train registered nurses in echocardiography for screening has been conducted in Fiji. In this model, nurses received a short period of training and undertook focused echocardiography, with a conservative threshold for referral to expert review. This approach appears to show promise in resource limited settings.[291]

Practical issues for echocardiography screening programmes

Even when echo screening is being used for epidemiologic research, providing ethical and appropriate clinical care is essential. A number of logistic issues should be considered.

Technical specifications of echocardiography machines Echo machines for screening require basic two dimensional and colour-Doppler imaging. Battery life or power source should be sufficient to maximise screening time each day. Plans for recharging batteries and backup power supplies should be established. The echo machine will need to be stored in a temperature stable, dry and dust free environment. Images from each day’s screening will need to be saved and can take some time to process. You should estimate data storage needs in advance, and ensure you have sufficient supply of CD’s or other storage devices.

You will also need to consider scheduling use of the echo machine which may be needed by a number of different services simultaneously, including pediatric clinics, adult clinics, community visits, visiting specialists and acute admissions.

Echocardiography screening programmes have predominantly used portable echo machines. The role of smaller handheld machines is the subject of ongoing research.[334]

Box 21: Considerations when purchasing an ultrasound machine for echocardiography

- Does it have two dimensional, pulse wave and colour Doppler imaging required to apply the WHF criteria?
- Can it store, save or transmit images?
- Is additional software required or included? e.g. Dicom
- 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 trans-oesophageal)
- 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 on bad roads, by boat or in dusty conditions?

Location of screening

Echo screening needs to occur in darkened rooms to improve image quality and interpretation.[335] Most screening occurs in schools and locating suitable rooms for screening and blackout materials may be challenging . Advance planning about the location and darkness of echo rooms is required. Where possible, rooms should be away from

electrical interference – including from mobile phones or power lines- which can distort echo images. Benches or plinths are generally required for people to lie on while being screened.[336] A reliable power source is required to recharge batteries. In some locations an electricity generator may be required to undertake remote field screening where power sources are compromised.

Expert review of images

Expert review of images is required for most programmes. However, large volumes of images can rapidly overwhelm specialist clinicians and create substantial delays. This time delay may make it difficult to find children who have been screened, and increase loss to follow up.

Privacy

In some countries and communities it is appropriate for young people to disrobe for echo screening. In other places, adolescents, particularly girls, will need to change into gowns. Adequate privacy for changing and screening should be arranged, and it may be necessary for female echocardiographers to screen young women.[336, 337]

Consent

Practices for the informed consent to screening of individuals or communities vary worldwide. Implications of screening may be significant in some locations, potentially affecting insurance status, marriageability or social status. Specific issues are best addressed by local or institutional ethics groups and this process should begin well in advance of any screening programme. Discussion about false positive and false negative results should also occur –there is good evidence that quality of life outcomes in screen positive- but not necessarily full echo positive- children and parents are impaired.[338] In many places written parental consent is required for echo screening – in these cases, ensuring that consent forms are provided for all the children being screened is critical.[336, 337] (See also discussion of consent, Chapter 23)

Recording outcomes

One of the big challenges in echo screening is managing crowds of interested children, and ensuring the process is as efficient and accurate as possible. Knowing who has consented, who is being screened and what the follow up is can be a challenge and requires significant administrative support.

Communicating results to patients and families

Afternoon education sessions were arranged in Nauru after screening to provide information to screen positive children.[339] In New Zealand, follow up letters are generally sent to primary care providers and to families, informing them of results.[336]

Box 22: Congenital heart disease

A background rate of congenital heart disease (CHD) occurs in children born around the world. Any screening programme to identify children with RHD is likely to identify children with previously undiagnosed CHD, some of whom will require operative management. For example, in a New Zealand cohort of 685 school children, 22 were newly diagnosed with CHD. This significant increase in cases was associated with a need for outpatient follow up and some operative intervention.[336] Programmes contemplating screening for RHD should anticipate new diagnosis of some CHD patients and have plans for their referral and management.

Box 23: Opportunity cost

The financial and human resources devoted to echo screening generally reduce capacity to deliver other components of RHD control programmes. For example, expert cardiologists may have to spend their time reviewing echo screening images rather than at scheduled follow up clinics for people already known to be living with RHD.

Case Study 7 | Rwanda

Dr.Joseph Mucumbitsi| President of the Rwanda Heart Foundation

The A.S.A.P. (Awareness, Surveillance, Advocacy, Prevention) Programme approach to RHD prevention is applied in a number of African countries. In Rwanda, one of the project demonstration sites is in the Gasabo district in Kigali. Dr. Joseph Mucumbitsi, founder and President of the Rwanda Heart Foundation and champion of the A.S.A.P. Programme in Rwanda, shared his experiences:

Awareness: "We want to focus on health worker training and get into schools." At the demonstration site in Gasabo, 10 schools were randomly selected, some urban and some rural, to take part in an echo screening campaign in conjunction with Team Heart, a surgical visiting team from the USA (see Case Study 8). The goal was to elucidate the prevalence of disease in the district as a first step in determining the burden of disease in Rwanda. At these schools, all students, teachers and a majority of parents were educated about RF/RHD, and RF/RHD posters were distributed. Nurses at the local health centres were trained in RF/RHD prevention and treatment. These nurses care for children from their catchment area schools. "We want the nurses at the health centres to be the ones that own the project… with education." A grant from the World Heart Federation has also funded the creation of education materials for RF/RHD, including a booklet for patients with RHD to streamline follow-up care, and a protocol for health workers on how to administer BPG, prevent endocarditis and monitor INR.

Surveillance: The WHF echocardiography criteria were used to diagnose RHD during the first echo based RHD screening campaign. 3,000 children, aged 6-16 years old, were randomly selected from the 10 schools. Plans are under way to expand the echo screening to another high prevalence region in the country to corroborate the initial findings. In the near future, 6 general practitioners' will enrol in a one-year Diploma in Cardiology including a course on echocardiography, so that local dependence on foreign experts to perform screening is reduced. The goal is to train GPs in cardiology from all District Hospitals so that they can help with CVD/RHD patients' diagnosis, management and follow up care under supervision by cardiologists, through a regular outreach programme.

Advocacy: "Our advocacy approach for CVDs in general and RHD in particular, started through the partnership with visiting cardiac surgery teams in 2006 aiming at building up a sustainable cardiac surgery programme. Two teams currently operate on RHD patients twice a year and so far, more than 250 patients have been operated on, allowing health care system strengthening for their follow up care." Due to the collective advocacy efforts of the RHD community in Rwanda, the A.S.A.P. RHD model components will be incorporated as much as possible into the national strategic NCD plan. Collaboration between the RHD community and the Ministry of Education is also promising, and progress is being made toward the inclusion of RHD in the national school health education programme. The school-based health care approach would be to ensure that a nurse from a local health centre is responsible for providing holistic care for students in neighbouring schools, including GAS pharyngitis treatment and referral of patients with symptoms of RF/RHD. Discussions with Partners in Health Rwanda, an NGO working with 3 District Hospitals in the country, resulted in switching from oral penicillin to injectable BPG for secondary prophylaxis. Discussions are being held with the national Treatment and Research AIDS Center to add RHD to 'TRACnet'. This would facilitate secondary prophylaxis follow up as well as the implementation of a web based central RHD register.

Prevention: Children who are echo-screen positive for RHD are started on monthly BPG injections. Borderline patients are not started on BPG because the natural history of asymptomatic and borderline disease remains unclear. There may be opportunities for Rwanda to become involved in international research to understand the role of secondary prophylaxis for borderline patients. Post cardiac surgery follow up care is provided through established "Points of Care" (8) throughout the country, where patients who have undergone surgical valve repair or replacement can receive BPG prophylaxis and anticoagulation management. At these sites, "Patients are seen by Cardiologists, General Practitioners or nurses and we (cardiologists) provide supervision through outreach visits, by phone or internet."

Challenges: There is no central registry of RF/RHD patients. Cardiologists keep spreadsheets of their own patients for follow-up purposes, but these are all maintained individually. The Rwanda Heart Foundation are still advocating for the Ministry of Health to endorse the idea of a registry, ideally integrated with an existing platform and web-based. Resources to expand the A.S.A.P. model and to achieve large-scale changes are limited. Sustained progress in RF/RHD control requires government commitment. Engaging government started in 2006 with the cardiac surgery programme supported by the Ministry of Health, and has strengthened since the creation of the National NCD Division including a CVD Unit. We have been working closely with them over the last year on the integration of RHD in the National NCD Strategic and Action plans.

"It's not just about what happens in the operating theatre, but it also involves good case audit, and more

Finucane and Wilson, New Zealand, 2013.[340]

Tertiary interventions (medical management of symptomatic RHD, anticoagulation, triage of intervention candidates and delivery of cardiac surgery) have not typically been included in RHD control programmes. There is often a distinction between local, register based, RHD programmes and advanced tertiary care delivered by humanitarian groups or visiting experts. It is true that these tertiary services do not have an impact the incidence of RHD, and will not control the disease at a population level. However, the individual burden of living with RHD can be reduced by access to tertiary services which can control symptoms and extend life. Traditional control programmes and interventional teams can both benefit from a collaborative approach to patient care and system strengthening.

The suffering of people with severe RHD provokes a strong humanitarian drive for clinicians and communities to find ways of accessing surgical interventions. Few endemic settings have access to local cardiac surgical services; humanitarian cardiac surgery visits or medical evacuation programmes are a more common model of service delivery. Even when national or regional services exist, they are often geographically remote from the most endemic communities. In low resource settings cardiac surgery programmes tend to be highly visible. The immediacy, visible results and powerful human stories surrounding surgery, often attract funding, media attention and community support. The profile and considerable expense of tertiary care has prompted legitimate concerns that surgical services may divert funds from cost effective register based programmes. The differences in geographic distribution, financial resources, and local ownership between RHD control programmes (focusing on primary and secondary interventions) and interventional services (focusing on surgery) sometimes make communication between these groups difficult. The distinction between control programmes and surgical programmes may be a missed opportunity for synergy and mutual benefit. Incorporating interventional services into RHD control programmes has a number of theoretical benefits:

Improve clinical care

- Maximise the benefit of surgery by ensuring the most suitable candidates receive intervention.
- Strengthen capacity for post-operative follow up, including the continuation of secondary prophylaxis and anticoagulation.
- Strengthen referral systems and handover between different levels of the health system.

Support health care staff

- Knowledge transfer between local health staff and national/regional/visiting/international staff about local practices, health services and approaches.
- Support training and capacity development.

Support sustainability

- Humanitarian cardiac surgical programs typically have far greater fundraising capacity than local, register-based RHD control programmes. Working together may make it possible to distribute these resources more equitably to support both treatment and intervention.

Comprehensive RHD control programmes offer an unprecedented opportunity for diagonal health system strengthening and integrative care (see figure 7, introduction). Interventions which can impact on the burden of disease at a population level should be prioritised; particularly, robust systems for secondary prophylaxis and strengthening of primary prevention. However, where resources or opportunity permit, it is reasonable to include tertiary medical and surgical services within the remit of RHD control programmes. This approach appears to be of value to patients, clinicians and communities; centers of excellence are needed to better understand models of best practice.

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?

How does your programme engage with traditional healers and beliefs?

Box 24: Acute surgery during an episode of RF

Some people with RF will have severe carditis which does not respond to medical therapy and requires urgent surgical intervention.[340] The proportion of individuals requiring surgery for RF appears to vary by setting. In countries with access to acute surgical intervention you will need to establish criteria for referral for urgent surgical consideration. This should be decided in consultation with a cardiothoracic surgical service.

Diagnosis of RF

Diagnosis of RF is difficult worldwide, and is particularly challenging in low resource settings.[94] There is no definitive diagnostic test for RF - instead a cluster of clinical and laboratory findings relate to the probability of disease. These signs, symptoms and results were codified into the Jones Criteria in 1944 to define diagnosis of RF.[341] The Jones Criteria have undergone a number of revisions since then to reflect changing needs of sensitivity and specificity.[21,342] The Jones Criteria and clinical details relevant to the diagnosis of RF appear in Annex B.

Specialist review, by a doctor, paediatrician or cardiologist, is often needed to make a definitive diagnosis of RF.[343] 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. Your programme will need to develop protocols for the referral of suspected cases for specialist investigation.

Specialist evaluation should occur in the acute phase of the RF wherever possible. Some of the tests required for diagnostic confirmation (**ASOT titres** 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. This may require admission to hospital to observe symptoms and await blood test results.[29, 81, 261]

Management of RF

'Management of ARF primarily involves confirming the diagnosis, relieving the pain of arthritis and managing cardiac failure with medication or, rarely, surgery' [43]

You will need a local protocol for managing RF in the referral hospital or secondary setting. This provides the best opportunity to gather information for diagnosis, manage the acute episode and begin planning for follow up. Your management protocol will need to include a number of different areas, outlined in Appendix B.

Table 28: Potential health system roles for RF management

Primary Care	Secondary Hospital	Tertiary Hospital	Quaternary
Suspected case of RF identified • Referred for secondary evaluation • Register notified of suspected case	Admission and specialist evaluation • Definitive diagnosis made • Register notified of diagnosis • Referral to tertiary center if evidence of heart failure or complications	Admission for advanced medical management • Clinical management of heart failure • Referral to surgical center if required	Admission if acute surgery required

lve between the right atrium and the
ht ventricle.
eferrals*

- Notify the RHD register coordinator of a new person to receive secondary prophylaxis generally
- Collect and record as many contact details as possible, including cell phone number of families, usual village and key community contacts. Provide these details to the register.
- Contact the primary health clinicians
- Arrange a dental review where possible

regurgitation.

Table 29: Sample discharge considerations for RF
Australian Guidelines,[29] New Zealand Guidelines[262] and WHF RHD Curriculum[20]
*Seek consent where required or appropriate

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

Diagnosis and management of RHD

Heart valve damage from RHD causes permanent changes to the way blood is pumped around the heart. Over time, this abnormal heart function stops the heart from pumping properly. Once people are symptomatic, RHD typically causes progressive activity limitation and breathlessness from heart failure.

Heart failure is the predominant medical problem of RHD. In an Australian population, 27% of people developed heart failure within 5 years of RHD diagnosis.[31] In many settings, RHD patients present late in the course of their disease - 46% of first presentations were in class **NYHA** III/IV heart failure in Uganda,[344] 18% in Soweto, South Africa.[163] Medical management of RHD involves using medications to control symptoms, improve outcomes and reduce complications.

Table 30: Common valve lesions in RHD

	Regurgitation (Stretching 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. Approximately 90% of people with RHD have mitral valve involvement.[44]	Mitral regurgitation occurs when 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.[345]	Mitral stenosis generally develops in more advanced RHD and is often caused by persistent or recurrent inflammation of the mitral valve.[346]
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.
Pulmonary Valve Valve between the right ventricle and the pulmonary artery.	The pulmonary valve is very rarely damaged by RHD.	

The high rate of teenage pregnancies combined with an endemic prevalence of rheumatic disease in developing countries results in cardiac disease being one of the most important comorbid states during pregnancy"

Essop and Nkomo, Valvular Heart Disease in Africa, 2005.[348]

Medical admissions

People living with RHD may require admission to hospital. In some parts of the world, these admissions are a significant proportion of inpatient cardiovascular care. In some areas of Africa, cardiologists estimate that RHD accounts for up to half of all the adult and/or paediatric caseload.[268]

Medical admissions for RHD tend to be for the treatment of heart failure, with diuretics. Admissions may be prolonged; average length of stay is 11 days in Fiji,[160] 13(+/-7) days in Cameroon,[79] and 3- 4 weeks in sub-Saharan Africa.[61] In a Nairobi community 27% of admissions for heart failure were precipitated by inadequate drug therapy, demonstrating the importance of good medical management, medication adherence and education.[195]

Contraception

Women of child bearing age with RHD need accurate information about safe pregnancies, or avoiding pregnancy. Locally available family planning, with a low failure rate, should be offered.

Opportunities for research

There has been some research into the experience of living with heart failure in high income settings.[355] However, much less is understood about symptoms and management in other communities. Qualitative research has an important role in understanding these issues, and alleviating challenges wherever possible.

Is there reliable data that provides insight into the extent of disease in people who present for the first time and are diagnosed with RHD, and why people do not seek care earlier?

Box 25: Traditional practices

"Patients do not usually do well without definitive surgery and so patients and their

Traditional, faith-based or community healers are a significant part of health care 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, with a gift that is to be respected.

People living with RHD also seek out traditional healers and therapy. For example:

- In Samoa, nearly 10% of people identified the 'village healer' as their first intervention for sore throat.[204]
- In Cameroon 45% of people attending hospital with heart failure (predominately from RHD) had already consulted a traditional healer.[79]
- Traditional beliefs have also impacted on management of RF or RHD in Hawaii,[53] Nigeria,[349] Zambia,[350] and Rwanda[41] and are likely to be influential in many other settings.

Traditional therapy is frequently perceived as delaying diagnosis and treatment.[41, 53, 79] 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.

Mortality and palliative care

Even with best medical therapy some people with RHD will die of their disease. Death from RHD occurs more frequently, and at a younger age, in settings when resources are limited and medical care difficult to deliver. In some places, the average age of death from RHD is very young:

- In Ethiopia the average age of death from RHD was 25.89 years +/- 11 years. 70% of people died from heart failure.[7]
- In the Indigenous Australian population of the Northern Territory, median age of death from RHD between 1997 and 2010 was 22 years. Over the 13 years of the study 28% of people on the register developed heart failure.[30]

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.

"**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 of pain and other problems physical, psychosocial and spiritual."[351]

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.[352] Treatment for pain, nausea, constipation and anxiety may also be required.[352] 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.[353] Resources for delivery of community based palliative care are available online.[354] Your programme may also provide support for families affected by deaths from RHD.

Taifa Leo newspaper coverage of rheumatic heart disease for World Heart Day 2013, published Sunday October 13th.

ood pressure
sphygmomanometer (blood pressure machine)*
stethoscope*

Blood pressure (BP) 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. BP measurements are also necessary for evaluating adverse drug reactions which may be associated with BPG delivery.

For auscultation of heart murmurs.

Medical equipment for management of RF and RHD

Different equipment is required at each level of the health system. A selection of relevant medical resources is outlined below. You may want to consider which resources are available (or should be available) in primary, secondary and tertiary settings associated with your programme. These can be marked off in the right hand columns of the table below. An example of this approach can be found in the Partners in Health Chronic Care Integration Guide for Endemic Non-Communicable Disease.[181]

★ Opportunities for integration

*Items with an asterisk are included in the WHO PEN Package of 'essential technologies and tools for implementing NCD interventions in primary care.[356]

Scales* and tape measure	For monitoring heart failure, nutrition and calculating body mass index (BMI).			
X-ray	Chest x-rays can be helpful for monitoring congestive heart failure, but add relatively little value over and above 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 (see box, chapter 16). ECG may also be valuable in confirming the diagnosis of arrhythmias such as atrial fibrillation.	☐	☐	☐
Resuscitation equipment	Including a defibrillator* and access to adrenaline for managing anaphylaxis.	☐	☐	☐

Echocardiography machines

An echo machine can be a very valuable addition to RHD control programmes and is useful for:

- Investigating incidental murmurs
- Quantifying valve disease
- Triaging people for operative intervention
- Planning operative intervention
- Cardiology review of valve lesion progression
- Monitoring post-operative outcomes
- Screening to estimate the burden of disease

Regular echo is also a valuable addition to symptom history, particularly in settings with cultural or language barriers which can make interpretation of clinical history difficult.[30] Trans-thoracic echo is sufficient for the majority of these tasks. Planning operative intervention may necessitate trans-oesophageal investigation in tertiary / specialist centres.

In many places a shortage of qualified staff to use echo machines and interpret results limits their use. Ensuring that the workforce issues are addressed before a machine is purchased helps avoid expensive equipment sitting idle and potentially falling into disrepair. Technical factors to consider when purchasing or accepting donated echo machines are outlined in Chapter 20.

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?

22 Anticoagulation

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.[21]

People who have had mechanical heart valve replacements depend on effective anticoagulation for survival.[21,357] Delivered effectively, anticoagulation prevents thrombus (clot) formation which could cause a stroke or block a mechanical heart valve.[357]

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 programme.

Warfarin anticoagulation

Warfarin is a vitamin K antagonist and powerful anticoagulation medication. Metabolism of warfarin varies between individuals and is affected by diet, exercise 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 (**INR**) and adjust the dose as required. This is called **variable dosing**.

Variable dose warfarin, titrated to INR is difficult to manage - even in relatively high resource settings. Management requires a very high level of health literacy for 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.[358]

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

- In an Indigenous Australian population, one third of RHD patients on variable dosed warfarin had inadequate warfarin monitoring.[359]
- In another Indigenous Australian group the TTR was only 44.9%.[360]
- In Nairobi, 103 patients on warfarin following heart valve surgery were in the therapeutic range only 17.85% of the time.[361]

Inadequate INR monitoring is associated with very poor clinical outcomes.[362] Approaches to INR monitoring and warfarin anticoagulation are outlined in tables 32 and 33.

Other anticoagulants

Aspirin

Aspirin may be sufficient anticoagulation for some patients with heart failure or low risk atrial fibrillation. Your RHD management guidelines should include advice on when aspirin is an appropriate option anticoagulation. The Partners in Health guidelines from Rwanda are a good example[94]

Heparin

Heparin is a short acting anticoagulation medication which is administered as an injection (either as intravenous infusion or a low-molecular weight form as a subcutaneous injection). Heparin may be used to provide anticoagulation for pregnant women unable to take Warfarin.

New fixed-dose agents

New medications for anticoagulation, which do not require blood test monitoring, are being developed.[357] These new drugs are not currently evaluated for people with heart valve disease or replacement, and remain very expensive for the foreseeable future.

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 a 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.[368] Education considerations when initiating warfarin anticoagulation may include:

- Indication for anticoagulation
- Target INR
- Tablet colours and doses
- Date and location of next blood test

in the laboratory model, patients have a blood test taken, blood is sent to a laboratory, INR measured, results are provided to health professionals and then instructions are provided to the patient about the dose of warfarin to take.

- Signs of bleeding. Information about when and how to seek medical assistance
- What to do if a dose is missed
- What to do if other medications are started or stopped
- The importance of a regular diet
- Planning a pregnancy, becoming pregnant, contraception
- Informing others of anticoagulation prior to surgical, medical and dental procedures
- Advice on sporting participation

INR monitoring

The major issue is medication adherence with warfarin and monitoring of INR.[348, 357] The therapeutic effect of warfarin is measured using the international normalised ratio (INR). The target INR depends on the type of valve replacement, arrhythmia or other indication. The dose of the warfarin needs to be changed (titrated) according to the INR. This should be done in small, incremental changes. There are a number of models for monitoring INR and titrating warfarin dose outlined in tables 32 and 33.

Supporting adherence

Providing medication packaged with the required medication dose each day has improved anticoagulation adherence in a vulnerable urban population in the United States.[36] Patient held INR record cards may also be an opportunity to communicate the target INR, date of next test and required dose.[369] This approach is widely used in high resource settings and in the Pacific Islands.[180]

> "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 blocked valve because the anticoagulant levels were not controlled, or a stroke, because they did not take the anticoagulants at all"
>
> Ali, Rebuilding the RHD Program in Sudan, 2013.[212]

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. Point of care testing has been adopted in a number of low resources settings and has made it possible to decentralise INR monitoring to local centres.[94] These machines require occasional testing/calibration and ongoing supply of reagent cartridges. Recommended technical specifications of POCT machines are available online.

Table 33: Models of warfarin dose adjustment

Primary care anticoagulation	In places where primary care is delivered by medical staff, anticoagulation monitoring is commonly arranged through primary care.[360, 365] This allows primary care doctors to provide comprehensive care for a range of medical conditions, and maintain frequent contact with people needing close monitoring.[365]
Pharmacist led anticoagulation	Anticoagulation education, dosing and monitoring may be arranged through pharmacy services.[366] Strong pharmacy engagement appears to improve time in therapeutic INR range and minimise adverse anticoagulation events in comparison to standard primary care.[139, 366] Depending on the skill and resource mix of your setting it may be possible to develop an integrated anticoagulation programme with a chemist or pharmacy.

Things to consider

How does your programme manage the list of people waiting for surgery?

Valve replacement

Does your programme have a relationship with a regular surgical or interventional service?

How do you communicate with surgical providers regarding potential interventional 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?

23. Triage & pre-operative evaluation

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 lasts for life.[21]

Overall evaluation of patients is important, particularly if multiple valves require replacement. The many complications of anticoagulation therapy can be a problem in remote areas. Some patients manage anticoagulation reliably and should not be denied best treatment. Others find compliance very difficult"

McLean et al, Australia, 2007.[375]

Your programme will need a system to identify people who may benefit from surgery (surgical candidates), evaluate their suitability for interventions, prioritise who can receive limited surgical resources and manage waiting lists. Systems to ensure that people are medically, mentally and emotionally prepared for intervention 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 (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. A summary of these are outlined in table 34; issues are broadly similar for other heart valves. Detailed discussion with a cardiothoracic service is needed to explore interventional options in your setting.

Table 34: Overview of mitral valve interventions for 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[48, 346, 370-372] The procedure is technically more difficult than valve replacement, and particularly difficult in RHD compared with other causes of mitral valve damage.[340]
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,[166] and in the Indigenous Australian context.[48] The closed approach reduces costs and complications compared with open surgical repair, providing a safe and effective option for low resource settings.[166] However, a cardiac catheterisation laboratory is required to perform the procedure and few facilities exist in the areas of greatest need.

Pre-operative issues for the health system

Who should be referred for evaluation? When should this happen?

Where possible, early engagement with cardiac services is desirable, to aid decisions about the timing of surgery and avoid missing the 'window of opportunity' for intervention.[372, 373]

Considerations of who can be offered surgery will usually include patient factors and health system factors, including:[94]

- Capacity of individual patients to benefit from surgery
- Ability of the surgical team to undertake both complex and relatively straightforward cases
- Post-operative ward capacity
- Training needs of local surgeons
- Cost of surgery
- Access to required follow-up, including anticoagulation and secondary prophylaxis.[340,375]

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.[372] There are a number of detailed clinical guidelines to inform these discussions.

WHO offers some clinical and echocardiographic indications for surgical referral.[21] In Australia, all symptomatic patients with clinical CHF are considered for intervention.[373] In Rwanda, cardiac surgical section is co-ordinated nationally by the cardiac surgery programme director and colleagues.[94]

The use of a priority based care planning system 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 (refer to priority based care planning, Chapter 19). Political interference in triaging referrals is a challenge in some countries.[179] Transparent criteria for referral may help address this issue.

How many people will require intervention?

Planning for interventional care should include an approximate estimate of the number of people who may benefit from pre-operative assessment, and system capacity to deliver interventions. This will vary by setting, waiting list and the type of interventions available, for example:

- In a USA cohort between 1985 and 2003 7.1% of patients on an RF/RHD register required surgery.[377]
- In an Indigenous Australian population 18% of patients required surgery over 10 years.[378]

"Aboriginal patients' natural anxiety about travelling away from their home communities for treatment is compounded by their experience of witnessing others who go to hospital and either never return or come back in a fragile physical and emotional state."

Lawrence, Australia, 2009.[380]

Beginning the informed consent process

Informed consent exists primarily to provide security to patients through the provision of relevant information, and to protect people from harm and exploitation.[383] The process is also one that provides an opportunity to engage the patient in the process, clarify ideas and levels of understanding, and provide legal protection for the clinician.[383] However, the Western medical concept of informed consent can be confounded by cultural norms associated with identity, self and family/community decision makers. Literacy, traditional and religious beliefs in developing countries and within sub-groups in developing nations may also confound informed consent.[384] National or local guidelines for obtaining informed consent should be available in the majority of settings.

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.[302] The process of obtaining informed consent takes time and ideally begins long before the date of surgery. Discussions about what is involved should begin early, allowing individuals and families to make a meaningful decision about the pathway forward.

Valid consent requires an understanding of relevant information. 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.

Miscommunication can make it difficult to gain informed consent. Misunderstandings may reduce the quality of surgical outcomes, and create fear and anxiety for others who will need surgical interventions. Culturally sensitive discussions with appropriately qualified clinical staff provide an important opportunity to address some of these issues. Many patients will need support to travel from their home village to the surgical centre. Practical issues are addressed in box 27, Chapter 25.

Infectious disease	Evaluation for potential infectious diseases will vary by setting but may include testing for tuberculosis, HIV, hepatitis C and malaria.[20, 21]	☐	☐
Nutrition status	A person with a good nutritional status pre surgery will have improved post-surgery outcomes compared to a person who is under nourished.		

Pre-operative issues for individuals

People living with RHD and being triaged for intervention may well have other health conditions or comorbidities. A preoperative 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).[378] A sample of pre-operative considerations are outlined in table 35.

Yur programme will need to discuss with your surgical team(s) (local, international or visiting)the role of each of these preoperative 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 the local setting, rather than having patients travel to tertiary centres and then be identified as unsuitable surgical candidates.[340] Investigations to be arranged by respective teams may be marked in the right sided boxes in table 35.

Table 35: Sample pre-operative investigations		Referral Centre	Tertiary Centre
Echocardiography	Echocardiography data provides critical information regarding valve lesions, cardiac chamber size, left ventricular function and pulmonary artery pressure. Serial data will assist with determining the timing of surgery.[29] Information about preoperative left ventricular dysfunction also provides information for risk stratification, improving the information for consent.	☐	☐
Dental optimisation	Routine care plans for people living with RHD should include regular dental review. Dental optimisation prior to surgery is particularly important to reduce the risk of subsequent bacterial endocarditis.[380] In Australia, inadequate dental preparation was one of the reasons planned rheumatic valve surgery was postponed and the patient returned home.[381]	☐	☐
Pregnancy status	Female surgical candidates travelling for surgery should have their pregnancy status confirmed before departure. Pregnancy is not necessarily an absolutely contraindication to intervention but should be considered prior to travel. *"Some medical patients have been accepted, only to discover when they arrive that they are pregnant. Sadly such patients are returned home"* Abbass and Pearson, United States Affiliated Pacific Islands, 2013.[313]		
Routine pre-operative bloods	Liver function tests, creatinine, glucose, electrolytes.[94]	☐	☐

Things to consider

How do other local/visiting surgical services follow up patients in your setting?

How and when will responsibility for clinical care transition back to usual services?

How will post-operative patients be followed up for clinical, and outcome monitoring?

24. Post intervention review, follow up & audit

Post-operative outcomes for RHD interventions are variable worldwide. Surgical intervention outcomes are poor in many countries. Often this reflects difficulties following up patients, maintaining anticoagulation and identifying post-operative complications early.[373] Establishing a robust structure for follow up is important prior to delivering intervention services. This ensures that people receiving the intervention get the most benefit, that limited funding is used appropriately and intervention is delivered safely.

Follow up is important to optimise outcomes of individual patients, and critically important for outcome audits, which should be an essential component of all surgical programmes. Communication between the tertiary and primary health sector includes the use of defined care pathways pre and post operatively, and reduces post-operative complications that arise from ambiguity, and a lack of understanding relating to the ongoing needs of the patient.

Communication between the providers of tertiary care services should take into account the cultural aspects of the patient and the primary health workers who will be responsible for the ongoing management of the patient post-surgery, and between follow up reviews by cardiology teams. Effective communication begins prior to the patient journey to the facility for surgery, and needs to include the sharing of knowledge about the surgery, the prognosis following surgery, the need for ongoing secondary prophylaxis, the importance of other medications and routine reviews. This assists the patient to provide informed consent, and will develop a shared trust and understanding.[381]

Table 36: Models of care following intervention

Primary Care	Secondary Hospital	Tertiary Hospital
• Monitoring of complications • Repeat prescriptions	• Potential for 'step down' or convalescent care • Management of complications • INR monitoring	• Discharge education • Anticoagulation initiated if required • Follow up appointments scheduled

Box 26: 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 access the level of care required post-operatively in the local setting?

What care, structured pathways of care around the time of a procedure and safety of care provided which meets the needs of individuals, families and your local capacity for service delivery.

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 geographically remote patients to call with concerns?[385]

In the same way that CAV treatment guidelines and secondary prophylaxis guidelines help to standardise the care that patients receive, guidelines help to improve the quality and safety of care provided which meets the needs of individuals, families and your local capacity for service delivery.

A defined 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 ensure ownership and knowledge of the model, and should be reviewed at designated time periods.

In developing capacity at the local level, surgical centres can consider referral patterns of local health service providers and incorporate the local health services into the delivery of care. A significant part of capacity development to improve outcomes is the provision of education and training, and this can be based on recognised and agreed protocols, best practice guidelines and the use of structured care planning activities.

Box 27: Education is everything

2. Post-operative planning for individuals and their families

Supporting families

People who have had severe breathlessness and exercise limitation with severe RHD generally experience significant symptomatic improvement after surgery. In some cases this is misinterpreted as a 'cure' and patients may stop taking any kind of medication.[346] Communication with patients and families is essential to explain that surgery is not a definitive solution for RHD. Many post-operative patients will require lifelong anticoagulation, secondary prophylaxis and medical supervision of pregnancies. However, as described in Cameroon, "those parents are seeing their child healthy and they think that they don't need any medication anymore."[346]

People in hospital for interventions are likely to have a prolonged admission. In New Zealand, children having surgery for RHD had an average length of stay of 13.5 days. Children who required surgery during ARF stayed an average of 54 days (+/- 16).[386] Around the world, 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.

Post-operative planning should occur far in advance of surgical procedures, be addressed when informed consent is secured, be reinforced during the hospital stay and at every post-operative visit. Children's HeartLink have developed a PEDI (Parent education/discharge instructions) resource for delivering post-operative education in a variety of languages and low literacy settings.[387] These are complemented by a web-based training module on caring for children before and after cardiac surgery.

3. Post-operative planning for the surgical team

All surgical services should be able to measure post-operative outcomes in order to give accurate information for informed consent, ensure practice is consistent with local/international standards and to facilitate ongoing improvement. Follow up of surgical patients is often limited in low resource settings, particularly when surgery has been curative and patients can be discharged from follow up. However, given the ongoing review needed following RHD interventions, a high standard of outcome monitoring should be expected. Where possible, cases should be presented at regular institutional meetings and be open to peer review. A report on the provision of surgical care by international organisations in developing countries notes, "Although all aspire to provide high-quality care, outcomes evaluation should be an integral part of every programme, especially when services include invasive procedures with the capability to harm as well as help."[388]

High quality outcome monitoring has been possible in high resource settings, particularly the Pediatric Cardiac Care Consortium in North America.[389] Reports from some single centres in low resources settings have also been possible.[385] A number of programmes have identified indicators for monitoring and evaluation of surgical processes.[181] Guidelines for reporting standardised mortality and morbidity following cardiac valve interventions is applicable to adult and paediatric patients undergoing all types of procedures.[390]

Establishing a framework for ongoing audit – including standardised data collection forms and recall schedules – should be embedded into surgical planning. The collection of standardised data will assist with evaluating the outcomes of surgical interventions, improve the quality of surgical care and reduce the cost burden. Evaluation strategies should be standardised to ensure data from each time period to another is comparable. The evaluation data can be helpful when redesigning the RHD control programme, determining future budgets, demonstrating cost benefits and influencing public policy.

Data should include the spectrum of procedures performed, the number of people who have had surgery, surgical outcomes, post-operative complications, the availability of resources (including the resources that were not available) and estimates on the number of procedures that did not take place due to limitations in resources.[392]

"Audit processes and risk model development and assessment are an essential part of this complex surgical charity work and will result in improved patient selection and outcomes"

Davis et al, Cardiac Surgery in the Pacific Islands, 2011.[391]

"It is the responsibility of the operating surgeon and team to audit the outcomes of this type of surgery rather than assuming the outcomes will be similar to those in a first world situation"

Finucane and Wilson, Priorities in Cardiac Surgery for Rheumatic Heart Disease, 2013.[340]

Case Study 8 | Rwanda | Team Heart

Ceeya Patton-Bolman | Registered Nurse | Programme Coordinator

"Ethically, it is important to support screening and prevention programes, as well as surgical intervention. From the very beginning, prevention and early intervention was a part of our mission statement."

Formed in 2006 after an invitation from the Rwandan Ministry of Health, Team Heart, a not-for-profit organisation based out of Boston, USA, began providing surgical care for children, adolescents and young adults with RHD in 2008. The organisation simultaneously engaged the Ministry of Health, local physicians and other entities to encourage and support the implementation of national efforts to prevent RHD. Ultimately, the team's goal is to strengthen the Rwandan capacity to control RHD until self-sufficiency is attained—"The best thing that could come out of this is for surgical teams doing just patients with valve disease due to RHD to be put out of business." Ceeya Patton-Bolman, a registered nurse and Programme Coordinator, reflected on Team Heart's efforts in Rwanda:

Components of the Programme

"One of the most valuable things about cardiac surgery is that it receives a lot of attention. When we travel to Rwanda, people know Team Heart and it is easy for us to parlay that into an awareness platform."

In 2009 Team Heart organised with the local cardiologist an RHD conference in Rwanda, which brought together cardiologists, paediatricians, the Ministry of Health, and Ministry of Education. Widespread agreement of the need to develop a national school-based screening and prevention programme for RHD was shared amongst participants. A vision of a programme was designed to identify and empower school nurses to educate teachers and families, diagnose and treat GAS pharyngitis, and refer patients with symptoms of RF or RHD to appropriate centres.

In 2011, a school-based screening initiative of 3,000 children in 10 schools took place in conjunction with the Rwandan Heart Foundation and Ministry of Health as part of the A.S.A.P project. Its purpose was to establish disease burden and use the data to push an awareness, prevention, and early intervention agenda. Education campaigns have targeted different populations:

1. Physicians: An annual national conference was held in conjunction with visiting teams.

2. Public: Posters were placed in local health clinics throughout the country and public forums were held at schools with parents and teachers.

3. Patients: "We think that our post-op patients can be our best ambassadors." Each year post-op patients come together for a reunion where advocacy is emphasised. Post-operative patients are encouraged to speak about RHD in their communities and increase awareness about opportunities for prevention and treatment. Reunions also include counselling on reproductive health, education, and return to normal productive life and activity.

4. Media: Radio shows with local cardiologists and patients have been used to raise awareness. Films, including "Heart of Courage" were used for fundraising, but were also used as tools for education in Rwanda. A skit/drama is in production and will be aired by a local TV station. Translation into Kinyarwandan will be completed soon.

"*One goal is capacity building so that Team Heart is not necessary for screening… We have not reached this point yet and are still working in collaboration with local staff.*"

Team Heart designed a diploma curriculum taught by a local cardiologist and partner visiting faculty from the United States and Europe. The diploma will train internal medicine physicians in basic cardiology and echocardiographic screening and also in the management and monitoring of heart failure. The initial cohort of 6 physicians will begin the 2-year programme in 2014. Team Heart also plans to "take echo machines to patients" after obtaining a grant with funding for two machines.

Transport and traditional healers

"We had a young man walk for 6 hours to see us. He had no money for a taxi which would have cost $1.50 one-way. Many patients are subsistence farmers and to buy anything they barter their agricultural harvest. But, they probably need to also use it to buy cooking oil or some other item necessary to live on. They never have paper money so they can't spend money to take themselves, let alone the entire families, to the regional health centre. So, they first reach out to traditional healers."

Challenges

1) Lack of registry

"… [Secondary prophylaxis] is a point for concern. It is currently being managed by local cardiologists, but to this moment there is not a central registry in the country of patients with RHD despite multiple requests from many areas including the World Heart Federation, WHO, Ministry of Health and partner expatriate teams".

Local cardiologists have an enormous clinical burden, inadequate administrative support and have struggled with consensus on a register format, ownership and responsibility. Secondary prophylaxis is overseen by

cardiologists, but is dispensed at local health clinics where someone is needed to document patients' clinic visits and would need to communicate compliance. Resource constraints make it difficult to hire dedicated staff to manage the registry or track patients. After looking through patient files and realising that patients were not receiving secondary prophylaxis as needed, Team Heart recently implemented this aspect of a surgical registry to monitor post-surgical patients. The organisation stresses teaching with each patient to remind them that they should be on secondary prophylaxis for the rest of their lives. The success of an agreed registry is contingent upon community support.

2) Lack of human resources, high turnover and low availability of technology

"*There may be as many as 10 [echo machines] in the country but uniform data base is not yet established. I was recently touring in a new hospital with two new, beautiful echo machines, but no one was yet trained to use them.*"

There are only 12 paediatricians and 5 cardiologists, of which 2 are paediatric cardiologists, for a nation of almost 11 million people. Two are assigned to the public sector. Team Heart Missions are complicated not only by a shortage of health care workers, but also by high turnover and internal migration of workers throughout the country. This internal migration has hampered consistent training of personnel, particularly nurses.

"*Nurses from Rwanda may be moved around frequently so that someone that I was working with and made progress with in February may well have moved on to improve the skills of a new hospital when I return in July.*"

3) Funding

"*Many programmes are put on hold because we don't have the finances to facilitate (them).*"

Team Heart wishes to hire someone to support and manage a central registry for a local cardiologist, but have been unable to secure stable funding for a position. It will take an influx of $5 - 7 million to build a self-sustaining cardiac surgery programme and strong awareness and prevention programme in Rwanda. Currently, the organisation is exploring the possibility of forming a public-private venture to raise funds.

4) Poverty and health systems challenges

"You need a programme that is affordable to the most vulnerable populations because RHD is a disease of poverty and a disease of childhood. Both are populations that do not consistently get a voice at the table."

From symptoms to systems

"*Most patients with newly symptomatic heart failure present to very rural health clinics that are staffed by nurses. That child is then totally dependent on the nurse having knowledge to recognise strep throat, RF/RHD symptoms. The nurse must then refer the patient on to the next health centre which is a distance away and requires money to get there. At that health centre, patients are further passed on to one of three referral hospitals where they would see a cardiologist for the first time. At that point, the cardiologist does a very thorough work-up. If they have an echo machine they will use it; if they don't the patient will be referred to yet another centre with a functioning echo machine. Then that patient will be placed on a list - the national list for cardiac surgery. Later, when one of the two foreign surgical teams doing rheumatic heart disease surgery comes into the country, they first look at that list and those patients are evaluated for surgical care.*" A small number of patients will be transferred for care abroad to India, Sudan, or Israel.

Provision of interventional services

Usual location of the surgical team

Local or national

International

diac surgical care

ntries with a high burden of RHD have
local access to surgical interventions for the disease.[392,393]
A variety of approaches for delivery of surgical services have evolved to address this unmet need.[166, 174, 394-397] These models are summarised in table 37 and addressed in more detail in the following sections.

Table 37: Models of surgical care

	Location of the procedure	
	Local or national	International
	National /regional centers of excellence This approach requires a sustained effort from local and regional agencies for the creation and continued operation of such centres and for the ongoing training of the workforce in resource poor settings.[174]	**International surgical training** Some countries have programmes for local surgical staff to travel and receive international training. These staff are generally expected to return home to work with local and international teams, maintaining competencies and passing on skills.
	Humanitarian surgical missions In some resource limited settings 'fly in fly out' teams are an important component local health services. At least 84 different organisations are providing paediatric cardiac surgical missions.[398] The number of adult providers is unknown. Humanitarian cardiac service delivery is supported by a number of international organisations.[399]	**Surgical evacuation** In some countries patients must travel internationally for operations. This may be arranged formally through the health system or privately by individuals or families. Follow up generally occurs in the patient's home country.

National or regional centers of excellence

Ideally, cardiac surgery should be delivered in settings which are geographically and culturally close to countries with a high burden of RHD. Although the development of cardiothoracic services in low and middle income countries remains challenging, promising models of service delivery have developed over some decades.[400]

In the African continent, independent cardiac services exist in South Africa, Egypt, Sudan, Kenya and Namibia.[400] In Cameroon, a regional centre hosts visiting surgical teams while local cardiologists undertake consultations, diagnosis, pace-maker implantations and cardiac catheterisations.[401] Similarly, the Salam Center for Cardiac Surgery in Sudan has provided cardiothoracic services since 2007 with a local and Italian team.[402, 403] The National Cardiothoracic Service of Ghana has been operational since 1992 with care delivered to patients from throughout the West African sub-region.[399]

Considerations in these settings may include:

- Supporting the training required for surgeons to develop valve repair techniques. The need for extensive personal experience may make it necessary for a single trainee to focus on repair skills.[340]
- The volume of patients required to develop and maintain surgical competency.
- Economic effects of a local surgical service, including potential cost savings,[399] or the sustainability of charitable financial support.[404]
- The opportunity cost (see box 23, Chapter 19) of a cardiothoracic center versus other investments in health.
- Retention of trained staff.

Visiting humanitarian surgical missions

While there are considerable benefits from international cardiac surgery missions there are also enormous challenges, and significant potential for harm. For example, in 2008 a visiting surgical team from New Zealand travelled to Samoa to provide heart surgery. Fourteen operations were performed- 13 for rheumatic heart disease- 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.[72, 406, 407]

Case volume and experience are significant determinates of surgical proficiency and outcomes: rheumatic valve surgery is no different.[373, 376, 408] International teams should ensure that they have sufficient surgical experience to deliver the planned schedule of interventions safely. Robust plans to transition care back to local staff are needed. The relationship between visiting teams and local staff demands careful and continuous attention. Disputes and disagreements can damage programme continuity, clinical care and community confidence.[409, 410]

Ethical considerations for international surgical missions have been developed.[412] Training local staff should be emphasised wherever possible, and formal strategies for knowledge transfer established.[174]

ox 29: Logistics of international surgical
ransfer

nternational surgical transfers are an expensive and omplicated process. Many arrangements fall outside he usual scope of experience for the medical system. sues to consider may include:

Does the patient have a passport? Is other documentation required to travel?

Will family members be able to travel with them?

Will children and young people traveling alone be safe?

What kind of language translation facilities will be available?

Will pa
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International surgical training

International placements to develop surgical skills in high resource settings play an important role in transferring skills to new surgeons and teams.[490]

Challenges include:

- Supporting surgical staff from low resource settings to return home following international training.[413]
- Sufficient case volume to acquire surgical skill in RHD which are rare in high resource settings.

"Your decision as a foreign graduate to return home after completing cardiothoracic surgical training (in) a Western country is personal and no one can make it for you… Although the road to establishing a sophisticated specialty such as cardiothoracic surgery in a developing country may be bumpy and full of daunting twists and turns I am convinced that the goal is worth the effort"

Kabbani, Syria, 2011.[340]

International surgical evacuation

Health system supported

In some settings specialists visit for triage and follow up, while operations and interventions occur remotely. This model may occur within countries (Australia[377]) or between countries (Samoa[72]). In New Zealand, 50% of operations in the single paediatric cardiothoracic unit are on patients from overseas, particularly the Pacific Islands.[340]

Individually arranged

Triage and prioritisation may be limited. Access to surgery may depend on ability to pay or secure charitable funding. These patients are particularly vulnerable for being lost to follow up and having poor post-operative outcomes.

Annex A: Assessment

The TIPS approach emphasises the importance of consciously addressing each component of comprehensive RHD control programmes. Considering the activities in each domain makes it possible to identify areas which need to be strengthened or highlight successful components worthy of celebration. To support this process a TIPS Assessment Tool has been developed to help define and describe how each component of an RHD programme functions. Drawn from the chapters of TIPS, the assessment questions address burden of disease, infrastructure and service delivery components relevant to existing and emerging programmes. Working through the assessment may help to identify areas which would otherwise be overlooked or under-addressed during programme planning. This provides a valuable opportunity to consider the 'next steps' for programme development in your setting.

The tool was piloted in five sites in 2013 and revised to reflect the needs of a diverse range of settings. The TIPS Assessment Tool can be downloaded free of charge online from http://www.rheach.org/tips/

The TIPS Assessment Tool is descriptive, rather than analytic. However, using this standard framework to explore experiences it may be possible to better understand best practice in the future and to develop stronger recommendations for program delivery and implementation. With repeated use it may also be possible to understand programme evolution over time, and better understand the relationship between interventions and outcomes.

Annex B: Diagnosis and management of RF & RHD

on-steroidal anti-inflammatory drugs (NSAIDS) provide ery effective pain relief for painful joints associated ith RF. NSAIDS are so effective that early use can mask ne migratory polyarthritis that is typical of RF. The use f NSAIDS can make it difficult to accurately diagnose RF. lany programmes discourage primary care staff from sing NSAIDS until specialist review can be arranged.[29, 305]

Accurate diagnosis of RF is vital to minimise the number of false positives (people who do not have RF who are incorrectly diagnosed as having the disease) and false negatives (people with RF who are incorrectly diagnosed as not having the disease). The implications of incorrect diagnosis for individuals and the health system are outlined in table 36.[257]

Diagnosis of RF relies on the Jones Criteria, last revised in In 2002 the World Health Organisation proposed an update to the criteria – reproduced in table 38. The criteria have been further refined Australia to increase the sensitivity of diagnosis, particularly in recurrent episodes.[29, 257, 414]

Diagnosed disease status	RF	Not RF
Diagnosed RF	**True Positive**	**False Positive** • Individual exposed to the pain, inconvenience and potential harm of secondary prophylaxis without any clinical benefit. • Resources consumed providing unnecessary care.
Diagnosed not RF	**False Negative** • Missed opportunity to provide secondary prophylaxis • High risk of recurrent episodes of RF and subsequent heart damage	**True Negative**

Polyarthritis	Do not require any other criteria to be diagnosed as having rheumatic heart disease.
Erythema marginatum	
Sydenham chorea	
Subcutaneous nodules	
Recurrent attack of RF in a patient without established rheumatic heart disease.[b]	Two major or one major and two minor manifestations plus evidence of a preceding group A streptococcal infection.
Rheumatic chorea. Insidious onset rheumatic carditis.[b]	Other major manifestations or evidence of group A streptococcal infection not required.

[a] Patients may present with polyarthritis (or with only polyarthralgia or monoarthritis) and with several (3 or more) other minor manifestations, together with evidence of recent group A streptococcal infection. Some of these cases may later turn out to be rheumatic fever. It is prudent to consider them as cases of "probable rheumatic fever" (once other diagnoses are excluded) and advise regular secondary prophylaxis. Such patients require close follow up and regular examination of the heart. This cautious approach is particularly suitable for patients in vulnerable age groups in high incidence settings.
[b] Infective endocarditis should be excluded.
[c] Some patients with recurrent attacks may not fulfil these criteria.
[d] Congenital heart disease should be excluded.

	Arthralgia

Abbreviations: CRP, C-reactive protein; ESR, erythrocyte sedimentation rate. Source: [21]

Investigation of cardiac involvement

Management of joint pain

Management of heart failure

Echocardiography where available

ACE inhibitors for severe heart failure

Aspirin for arthritis or severe arthralgia[29, 81, 261]

Clinical observation	Examine the patient daily and observe for clinical features of RF – see Annex E. Temperature, pulse, respiratory rate and blood pressure four time daily, including a sleeping pulse during the night and if pulse is greater than 100 beats per minute complete apical heart rate
Blood tests	• Inflammatory markers • Evidence of GAS exposure
GAS eradication	Antibiotics to ensure GAS is eradicated from the upper respiratory tract29, 261
Management of fever	• Low grade- does not require specific treatment[29, 261] • If required, fever on its own or with mild arthralgia may be treated with paracetamol[29, 261] • Fever will respond well to aspirin therapy[29, 261]
Management of arrhythmias	Digoxin where atrial fibrillation is present

Box 31: Sydenham's chorea

One of the unique manifestations of RF is Sydenham's Chorea – a triad of altered mood, muscle weakness and uncontrollable discoordinated movements. This chorea is most common in adolescent women.367 A major manifestation of RF and often associated with carditis,[367, 369] therefore echo is an essential investigation for patients presenting with chorea.[29] Patients with a history of chorea are considered to be at risk of subsequent cardiac valve damage and should be carefully followed up to ensure they receive secondary prophylaxis.[29]

Because chorea can present as a symptom of RF following a long latent period evidence of preceding GAS infection to confirm diagnosis is not necessary in some jurisdictions, once other causes of chorea are excluded. 29, 81, 261, 364, 367

Indicators of chorea include:[29]

- the 'milkmaid's grip' (rhythmic squeezing when the patient grasps the examiner's fingers).
- 'spooning' (flexion of the wrists and extension of the fingers when the hands are extended).
- the 'pronator sign' (turning outwards of the arms and palms when held above the head).
- inability to maintain protrusion of the tongue.
- Symptoms disappear when the patient sleeps.

Treatment:
Treatment is rarely indicated in most instances as the condition is self-limiting within weeks, often up to six months. The condition can be quite distressing for the person and their family; however there is no evidence that lasting neurological damage occurs. Reassurance and support for the patient and family are important, coupled with regular messages regarding the need for secondary prophylaxis to prevent further episodes. If treatment is necessary, the Australian and New Zealand RF/RHD guidelines recommend carbamazepine or valproic acid.

Annex C: Anaphylaxis

Anaphylaxis is highly likely when any one of the following three criteria is fulfilled:[279]

1. Acute onset of an illness (minutes to several hours) with involvement of the skin, mucosal tissue, or both (eg, generalized urticaria, itching or flushing, swollen lips-tongue-uvula)

AND AT LEAST ONE OF THE FOLLOWING:

A) Respiratory compromise (e.g. dyspnea, wheeze-bronchospasm, stridor, reduced PEF, hypoxemia)
B) Reduced blood pressure or associated symptoms of end-organ dysfunction (e.g. hypotonia collapse, syncope, incontinence)

OR

2. Two or more of the following that occur rapidly after exposure to a likely allergena for that patient (minutes to several hours)

A) Involvement of the skin-mucosal tissue (e.g., generalized urticaria, itch-flush, swollen lips-tongue-uvula)
B) Respiratory compromise (e.g., dyspnea, wheeze-bronchospasm, stridor, reduced PEF, hypoxemia)
C) Reduced blood pressure or associated symptoms (e.g., hypotonia collapse, syncope, incontinence)
D) Persistent gastrointestinal symptoms (e.g., crampy abdominal pain, vomiting)

OR

3. Reduced blood pressure after exposure to known allergenb for that patient (minutes to several hours)
A) Infants and children: low systolic blood pressure (age-specific) or greater than 30% decrease in systolic blood pressurec
B) Adults: systolic blood pressure of less than 90 mm Hg or greater than 30% decrease from that person's baseline

PEF: peak expiratory flow.
a Or other trigger, for example, immunologic but IgE-independent, or nonimmunologic (direct) mast cell activation.
b or example, after an insect sting, reduced blood pressure might be the only manifestation of anaphylaxis; or, in a similar example, during allergen immunotherapy, after injection.
of a known allergen for that patient, generalized urticaria (only one body organ system affected) might be the only initial manifestation of anaphylaxis.
C Low systolic blood pressure for children is defined as less than 70 mm Hg from 1 month to 1 year, less than (70 mm Hg 2 age) from 1 to 10 years, and less than 90 mm Hg from 11 to 17 years. Normal heart rate ranges from 80–140 beats/min at age 1–2 years; from 80–120 beats/min at age 3 years; and from 70–115 beats/min after age 3 years. Infants are more likely to have respiratory compromise than hypotension or shock, and in this age group, shock is more likely to be manifest initially by tachycardia than by hypotension.

ometimes called colonisation)

(quality-adjusted life years) can complicate the bacterial diagnosis of pharyngitis because there is no mechanism to distinguish carriage from active infection.[218]

Glossary

Aetiology	The cause of disease
Anaphylaxis	Anaphylaxis is a severe, life-threatening, generalised or systemic hypersensitivity reaction
Antibiotic resistance	The evolved insensitivity of a bacterium to a drug that it was once sensitive to.
Anti-DNase B	anti-deoxyribonuclease B An antibody produced in response to GAS antigens
ARF / RF	Acute rheumatic fever / rheumatic fever
Asymptomatic infection	The subclinical colonisation of host tissue by microbes.
BPG	Benzathine Penicillin G, also known as benzathine benzyl penicillin. A form of penicillin delivered by intramuscular injection producing prolonged serum penicillin levels
Cardiac auscultation	Listening to the heart with a stethoscope
Chorea (Sydenham's Chorea)	Sydenham chorea is a neurological disorder of childhood resulting from GAS infection. Chorea is characterised by rapid, irregular, and aimless involuntary movements of the arms and legs, trunk, and facial muscles. It may also be associated with erratic emotions and muscle weakness.
Conflict of interest	A conflict between the private interests of an individual or organisation and the responsibility of an individual or organisation in a position of trust.
Congestive heart failure	A condition in which the heart's function as a pump is inadequate to deliver oxygen rich blood to the body.
Continuous Quality Improvement (CQI)	The process of continuously monitoring the quality of services provided and using a structured approach to systematically improve these services.
CRP	C-reactive protein. Blood test used clinically as a non-specific marker of inflammation.
Denominator	The size of the target population, used as the bottom number of a fraction.

[illegible]

marker of inflammation.

tions on a larger scale.

Emm type	A way to identify different strains of GAS by antigenic variations in the N-terminus of the M protein.
Echo / Echocardiography	Echocardiography is a diagnostic test that uses ultrasound waves to create an image of the heart muscle.
Fact sheet	A short summary of important information
Focus group	An interactive group setting in which individuals are asked about their beliefs, experiences and attitudes towards a product or programme in order to improve the product or programme. Focus groups are an example of both qualitative research and feedback.
Health literacy	The cognitive and social skills which determine the motivation and ability of individuals to gain access to understand and use information in ways which promote and maintain good health.
Hypersensitivity	An immune-mediated reaction to an allergen...
Indirect maternal mortality	Deaths resulting from previous existing disease, or diseases that developed during pregnancy, and which were not due to direct obstetric causes but aggravated by physiological effects of pregnancy
IM	Intramuscular An injection of medication delivered into muscle.
Low and middle income countries (LMICs)	Low and middle income countries as based on Gross National Income (GNI)
NCD	Non-communicable disease
Non-governmental organisations (NGOs)	A non-profit, voluntary citizens’ group which is organised on a local, national or international level
NYHA	New York Heart Association classification of heart failure.
Household overcrowding	Too many people for the space or resources available. A number of criteria for defining and measuring overcrowding exist.
Pharmacovigilance	Detection, assessment, understanding and prevention of adverse effects or any other drug-related problem.
Position statement	A formal declaration delineating support, opposition for a policy or intervention or a statement of recommendations meant to provide direction for the target audience
Prevalence	The number of cases of a disease in a population at one moment in time

[illegible] methods	[illegible] statistical and mathematical data to measure phenomena.
Primary Care	The medical care a patient receives upon first contact with the health care system, before referral elsewhere or to a specialist.
Push/pull factors	Push factor: a factor that increases the likelihood of emigration. Examples include poor wages, restricted professional opportunities, and safety and security concerns, among others. Pull factor: a factor that increases the likelihood of immigration. Examples include good wages, opportunities for professional development, political stability, among others.
Quality improvement	In medicine, often refers to the process of overcoming the logistical challenges that prevent the delivery of evidence-based medicine for patients in order to improve patient outcomes while lowering health care costs
Recall bias	Error that results from study participants' inaccurate recollection or reporting of events from the past
Research questions	A question identified by an investigator which will be explored systematically and scientifically
Sensitivity	Measures the ability of a diagnostic test to accurately identify disease-positive patients. Defined as true positive results over true positive + false negative results. Se = TP/(TP+FN)
Social media	Virtual forums in which members share, create and exchange information and ideas.
Stakeholders	Individuals, organisations and /or governments that have an interest in the outcome of a policy debate or in the implementation of a programme plan.
Support groups	Groups composed of individuals with similar afflictions who are able to understand and relate to each other and thus provide empathetic support
Sustainability	The ability of a project to remain viable in the future with little to no further intervention
Triage	The act of categorising patients into groups based on risk or severity of disease, usually to prioritise access to medical care and resources.
WBC	White blood cells. Cells of the immune system circulating in blood Clinically, a high "white count" implies infection.
WHO	World Health Organisation

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