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RHEUMATIC HEART DISEASE

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Tools for implementing rheumatic heart
disease control programmes

‘QUICK TIPS’



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This 'Quick TIPS' is a summary of the full publication 'Tools for Implementing RHD Control Programmes (TIPS) Handbook'. References, annexes and a glossary are provided in the full handbook edition.

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 exact 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). 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. Robust epidemiologic data for RF and RHD is insufficient; the true burden of disease is likely to be several times higher than current estimates. Approximately half a million people die of RHD annually around the world. Overwhelmingly these deaths are premature; on average, people dying from RHD are aged under 40.

The majority of people with RHD live in developing countries. 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. 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. 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 and 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. 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%. For individuals with a history of previous RF the risk rises to 50%. The most important determinant of disease progression appears to be the number of times RF recurs in an individual. 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.

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

Register-based programmes for RHD control have been recommended by the World Health Organization (WHO) and World Heart Federation for many years. 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.

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. The A.S.A.P model incorporates four key elements: education, primary prevention, secondary prevention and disease surveillance. These components offer a clearly articulated policy overview of the domains required for disease control.

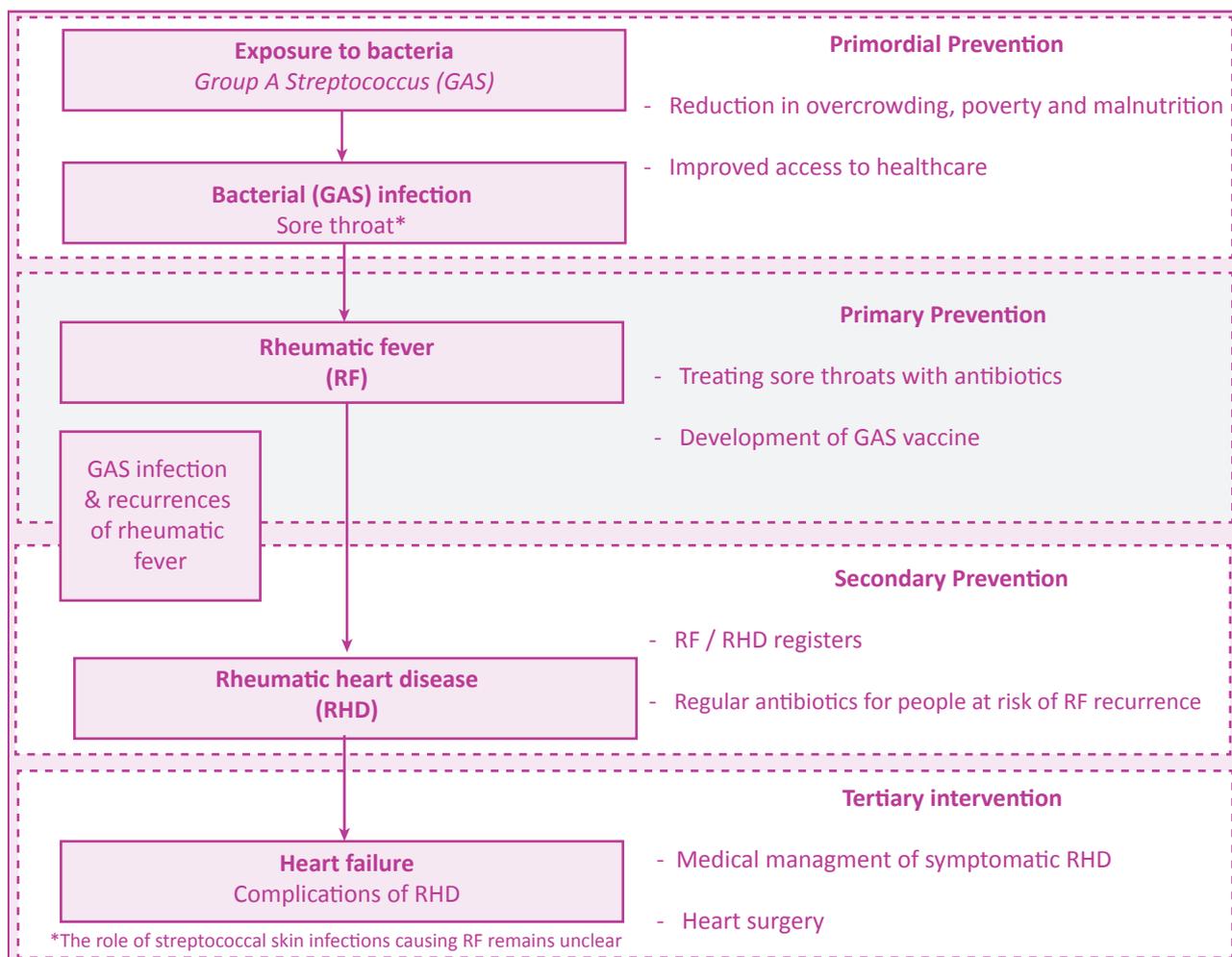


Figure 1: Causal pathway of RF/RHD and opportunities for intervention.

What is a comprehensive RHD control programme?

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. 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 2. The components are arranged in approximate order of priority, working from left to right, bottom to top, in each row. 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.

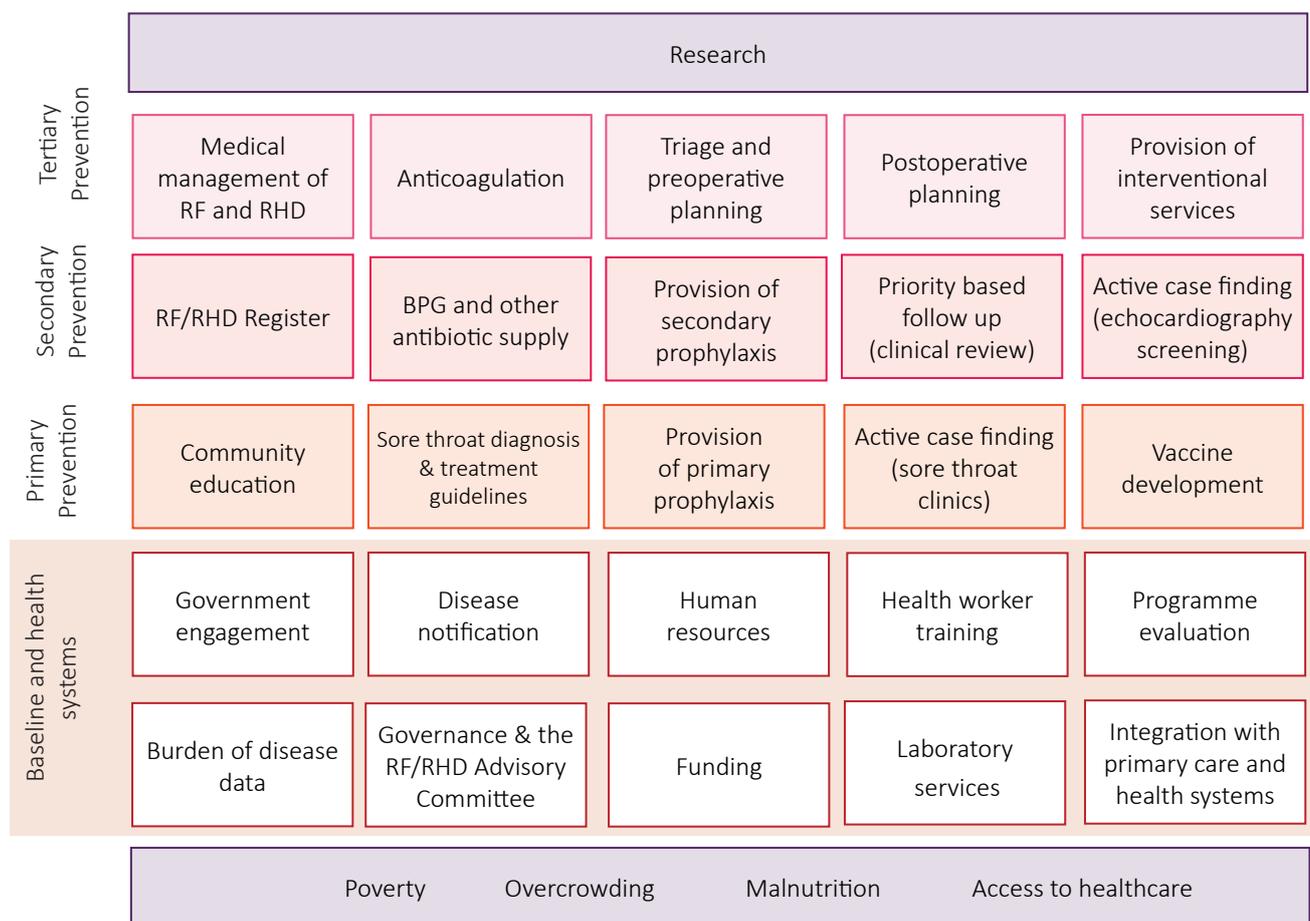


Figure 2: A conceptual framework for comprehensive RHD control programmes. Components are arranged in approximate order of priority, working from left to right, bottom to top, in each row.

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.

Health systems & baseline components

Baseline components should be considered (but not necessarily completed) before beginning an RHD control programme. They remain important throughout the duration of the programme and they provide a strong foundation for planning, implementation and evaluation of programme outcomes.

Successful RHD control programmes do more than simply deliver clinical care. Control programmes must interact with communities and the wider health system to provide effective prevention and treatment of RF and RHD. These collaborations are likely to be needed over many years before the burden of disease is significantly reduced. Robust programmes have a strong foundation of planning and strategy to support partnerships and deliver sustainable services.

Estimating the burden of disease is a critical starting point for many programmes. Burden of disease information helps your programme provide interventions which are focused in the areas of greatest need and to allow monitoring of your impact over time. The information is also useful for decision makers in government, funding agencies and for communities. Responding to this burden of disease information requires good governance, sustained funding, human resources and a structure for evaluation. Most programmes work with other parts of the health system; laboratories, training providers, primary care structures and disease notification measures to delivery prevention and treatment interventions for RHD.

The baseline and health systems section of TIPs outlines a number of these domains to consider when developing an RHD control programme. It is unlikely that your programme will be able to comprehensively address each area before delivering services – however each should receive conscious consideration and have a plan to strengthen and refine these components as the programme develops. Baseline components provide a strong foundation for planning, implementing and evaluating the outcomes of your RHD control programme.

1. Burden of disease data

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 so that it can be replicated in the future?

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

A clearly defined denominator is important for understanding the burden of disease in your setting. 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.

It is important to be aware that 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. The needs of vulnerable populations should be considered when collecting data and developing burden of disease estimates. Similarly, RF and RHD have a relatively predictable age distribution worldwide. 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.

Table 1: Sources of burden of disease data

	Advantages	Disadvantages
Hospital records	<ul style="list-style-type: none"> • Admission and discharge routinely collected in most settings. • Injection books or other records of care delivery may be accessible. • Valuable source for estimating the costs of care for RHD. 	<ul style="list-style-type: none"> • May underestimate the burden of disease because only the sickest patients present. • May overestimate the burden of disease because people from a large area attend tertiary centers for care, so some cases not from the same catchment area as the denominator may be included inadvertently.
Death records	<ul style="list-style-type: none"> • Routinely collected in most countries. 	<ul style="list-style-type: none"> • Variable accuracy depending on local standards of recording. • May be difficult to access.
Extrapolating from similar countries	<ul style="list-style-type: none"> • Can provide an approximate burden of disease estimate at low cost. 	<ul style="list-style-type: none"> • Imprecise. • Unable to monitor change over time.
Disease notifications	<ul style="list-style-type: none"> • Can provide high quality data with lots of clinical information. 	<ul style="list-style-type: none"> • Requires a central notification system and lots of education of the health workforce.
Echocardiographic screening	<ul style="list-style-type: none"> • Highest quality biological data about burden of disease. <p>*There may be an important role for baseline echocardiography screening when programmes are being established. A known denominator and standard diagnostic criteria are important – See Chapter 20.</p>	<ul style="list-style-type: none"> • Expensive to run echo screening. • May be unethical to screen unless clinical care can also be provided. • May overestimate burden of mild disease and include subclinical disease of uncertain significance. • May underestimate the burden of severe disease if school children are absent during screening

2. Governance and the RHD Advisory Committee

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, timeframe and terms of reference?
- What are their primary goals for the programme?

Designing and developing an RHD control programme requires input from many different stakeholders. 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. 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. Once programmes are established the Advisory Committee can offer valuable clinical, implementation and evaluation support. 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.

Table 2: RHD advisory members and stakeholders

Government	Health Sector	Community	Non-Government
<ul style="list-style-type: none"> • Ministry of Health • Ministry of Education • Ministry of Housing 	<ul style="list-style-type: none"> • Primary care/general practice/family medicine • Nurses/midwives and local health workers • Medical Specialists: <ul style="list-style-type: none"> • Cardiologists • Paediatrics • Internal medicine • Cardiac surgeons or surgical programmes • Dentists • Epidemiologists • Microbiologists • Laboratory services • Echocardiographers 	<ul style="list-style-type: none"> • People living with RF / RHD • Community leaders • Faith based groups • Family groups • Schools and teachers 	<ul style="list-style-type: none"> • World Health Organization • Donors and funders • National/international heart networks and societies • Private sector partners • International technical advisers

Table 3: Potential Roles of the RHD Advisory Committee

Clinical leadership	<ul style="list-style-type: none"> • 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
Setting strategy and goals	<ul style="list-style-type: none"> • Identify gaps in local data, gather or support research to fill them • Identify goals and targets for the programme • Track activities and data to guide the priorities of the control programme staff
Representing the programme	<ul style="list-style-type: none"> • Present the work of the programme at meetings, in the media and to the community
Engaging the community and other stakeholders	<ul style="list-style-type: none"> • 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.
Resource mobilisation	<ul style="list-style-type: none"> • Fundraise, mobilise resources or advocate for financial support of the programme
Evaluation oversight	<ul style="list-style-type: none"> • Provide programme governance, including sourcing internal and external evaluations.
Advising and mentorship	<ul style="list-style-type: none"> • Support individuals and programmes to expand RHD control activities in other locations

3. Funding

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?

Table 4: Preparing for fundraising

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. 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.
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.

Table 5: Potential sources of funding

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.
Development Assistance	Some governments have funded international control programmes and humanitarian surgical missions. Often these funds are arranged at a government level and require diplomatic collaboration.
Professional Organisations	Professional groups, including medical associations or professional colleges, can be instrumental in generating or helping to secure funds supporting project activities.
Charitable Donors	A variety of charities have donated funds or resources to RHD control programmes. These valuable contributions often support small local initiatives. Community organisations, faith based groups, business foundations and non-government organisations (NGOs) may prove to be valuable partners for your programme.
Businesses	Local (or international) businesses can sometimes be encouraged to donate funds, often for specific pieces of equipment. Businesses may also be willing to contribute their own products.
Community Fundraising	In many countries, families or individuals seek private support for private or international cardiac surgical interventions. Fundraising generally happens with events, through media appeals or via social media. You should consider how your programme will respond to these efforts - personal appeals may be a good opportunity to raise awareness about RF and RHD but it can be difficult to ensure that information is accurate and appropriate.
Research Funding	Appropriately, many RHD research projects in low resources settings have a service delivery component. Any research conducted in conjunction with your programme should address the clinical needs of people and communities living with RHD.

4. Laboratory services

Things to consider

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

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.

Table 6: Laboratories

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. This laboratory can provide, <ul style="list-style-type: none"> • Reference strains of GAS to ensure standardised results. • Expert advice on laboratory standards and training. • Specialist knowledge on sample testing and result interpretation.

Table 7: The role of laboratory facilities in RHD control programmes

1. The role of the laboratory in primary prevention of RF Supporting the diagnosis of primary GAS infection for primary prevention.

Throat swab cultures	Rapid antigen detection tests
Bacteria (including GAS) that are causing infection of the throat can be grown in the laboratory from bacterial swabs of the throat.	RADT tests are easy to use, commercial kits that detect specific parts of the GAS bacteria (antigens).

2. The role of the laboratory in diagnosis of RF Providing evidence of preceding GAS infection. Providing evidence of systemic inflammation.

Evidence of preceding GAS infection	Tests for inflammation and/or infection
GAS infection results in the production of antibodies specific to antigens expressed by GAS. 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.

3. The role of the laboratory in management of RHD and pre-operative evaluation Information for assessing potential surgical candidates and monitoring anticoagulation

Pre-operative evaluation	Anticoagulation
Laboratory tests for potential surgical candidates may include a full blood count, liver function test, creatinine, glucose, urea and electrolytes. Screening for infectious diseases may include tuberculosis, HIV, hepatitis C and malaria. Women with RHD also require access to pregnancy tests.	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.

4. Burden of disease Monitoring the epidemiology and serotype distribution of GAS within a population.

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. *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.

5. Integration with primary care & health systems

Things to consider

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

Table 7: Opportunities for integration

Sector	Rationale for integration	Opportunities for integration
Primary care	The principles of community based programmes transcend disease specific issues and focus on the needs of individuals and their families. Services can be affordable, sustainable and accessible to people living with RHD and other conditions.	<ul style="list-style-type: none"> • Support primary care clinicians to treat sore throats appropriately with local guidelines. • Empower the primary care system of to deliver or oversee primary and secondary prophylaxis.
Perinatal care	Women with RHD are at risk of illness or death during pregnancy and delivery. Worldwide RHD is a significant contributor to maternal death.	<ul style="list-style-type: none"> • Inform women with RHD about the need for medically supervised pregnancies and deliveries. • Train midwives and health workers to ask about a history of RF/ RHD and anticoagulation during antenatal screening. • Support access to contraception.
Non communicable disease	RHD programmes are often managed or administered by NCD departments. A chronic disease approach reflects the long duration of RHD and the inclusion of RHD in the WHO Global Action Plan for reduction of premature mortality from NCDs by 25% by 2025.	<ul style="list-style-type: none"> • Ensure that RHD is incorporated into national NCD strategic and action plans.
Congenital and other childhood heart disease	RHD and congenital heart disease can have similar symptoms and sometime similar medical and surgical treatment.	<ul style="list-style-type: none"> • Develop integrated support groups and education programmes for young people and their families living with RHD.
Health skin programmes	Although the association between GAS skin infection and RF remains unclear it is reasonable to consider including healthy skin programmes as part of RHD control.	<ul style="list-style-type: none"> • Ensure that hygiene education programmes include both skin sore management and reduction in RF risk.
Dental care	People living with RHD are at risk of infective endocarditis. Good dental hygiene reduces the risk of infective endocarditis.	<ul style="list-style-type: none"> • Support people with RHD access dental resources (toothbrushes and toothpaste) and clinical dental care.
School health services	Schools are an important opportunity for education about RF/RHD. It may also be possible to deliver services (primary and secondary prophylaxis) in a school based setting.	<ul style="list-style-type: none"> • School projects, posters, plays, lessons and community events

The relationship between streptococcal skin infections and RHD?

GAS pharyngitis causes RF and subsequent RHD. GAS (*Streptococcus pyogenes*) also causes the skin infection impetigo (pyoderma). Observationally there is an overlap of communities with a high burden of impetigo and a large number of people living with RHD. In particular, Australian Aboriginal communities demonstrate a high incidence of RF; in these settings anecdotal reports of sore throat are low, GAS throat carriage is very rare but impetigo is hyperendemic. This correlation supports the idea that streptococcal skin infections, in addition to GAS sore throat infections, may also cause RF- or perhaps make individuals more likely to develop RF following GAS pharyngitis. Conversely, in some Aboriginal communities, early studies found that throat carriage was not low.

6. Government engagement & advocacy

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?

Governments are one of the most important stakeholders in RHD control. 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. However, engaging government often depends on local custom, politics and government structure. Approaches to government are best guided by experience in the local setting

Table 8: Steps to effective advocacy

1. Know the issue
2. Establish goals
3. Identify the problem
4. Identify existing advocacy activities
5. Create a plan or campaign
6. Engage with appropriate elected officials
7. Meet with officials
8. Use communication tools to strengthen messages
9. Track success and share outcomes

Table 9: Opportunities for government engagement

	Rationale for engagement	Individual roles	Integration
Ministry of Health	The Ministry of Health generally allocates or oversees resources for RHD control and establishes national disease control priorities and policy.	<p>Clinicians and professional groups</p> <p>‘RHD champions’ have been critical disease advocates in a number of settings. Worldwide, clinical advocates have been critical for maintaining RHD on the national and international agenda. Clinical organisations also have a credible professional voice to call for resources, attention and action to RHD. 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.</p>	<ul style="list-style-type: none"> • Align RHD control program priorities with national health plans. • Work towards common goals for disease control. • Undertake shared research projects. • Provide local data to support decision makers.
Ministry of Education	School age children are at greatest risk of RF, making schools a sensible target for disease intervention. RHD prevalence studies to define baseline burden of disease are generally undertaken in primary schools where a denominator is easily defined. Schools also provide an important opportunity to educate communities about RF/RHD.	<p>Teachers and educators</p> <p>Teachers may have an important role in identifying children with a sore throat, symptoms of RF (particularly joint pain and chorea) and children with heart failure. They may also be able to identify and assist children falling because of poor health and reduced school attendance.</p>	<ul style="list-style-type: none"> • Prevention programmes, care delivery and specialist outreach may also be integrated into school programmes. • Administrative or logistic support from schools to record secondary prophylaxis adherence, notify programmes of transfers or new students. • Delivering education and services through schools generally requires support from the Ministry of Education, and often at an individual school level.

7. Disease notification

Things to consider

- Are there notifiable diseases in your 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?

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. 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 10.

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.

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. 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.

Table 10: Criteria for notifiable conditions

A recognisable disease	RF can be diagnosed by the modified Jones criteria (see Annex B) although a different case definition may be used to trigger notifications
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.
There must be the potential for action	For individuals with RF, notification can be linked to a register based programme which provides secondary prophylaxis and prevents recurrences of RF.
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.
Notified data should be usable	RF notifications offer an opportunity to understand the distribution and burden of disease, plan interventions and monitor outcomes.

8. Human resources

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 co-ordination 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?

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.

Table 11: Tasks in RHD control programmes

Clinical	<ul style="list-style-type: none"> • Expert advice on guideline development and implementation • Consult and train primary health staff • Echocardiographers • Physiotherapy
Allied Health	<ul style="list-style-type: none"> • Laboratory staff • Epidemiology support
Administrative	<ul style="list-style-type: none"> • Maintaining the register- database developer and manager • Arranging Advisory Committee meetings • Arranging community education programmes

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.

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, BPG 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.

Strategies for retaining staff

The global health workforce shortage can make 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 12.

Table 12: Push & pull factors in health worker retention

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

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.

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. A plan for training

activities, expected competencies and outcomes should be developed. 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.

Communities with a high burden of RF and RHD also tend to be areas with a high turnover of health workers. This makes ongoing training - and the development of accessible resources - particularly important. In particular, new staff should be made aware of the symptoms of RF and the need for specialist evaluation to confirm diagnosis. Every episode of RF which goes unrecognised is a missed opportunity to begin life changing secondary prophylaxis.

Table 13: Health worker training

Dedicated RF and RHD training	Embed into existing training
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. However, bringing people together especially for RHD training can be expensive, and may interrupt the provision of health care in settings where human resources are limited.	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.

Table 14: Publications including RF and RHD

Dedicated RF / RHD publications	Embed in existing publications
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: Local guidelines/handbooks for the management of RF and RHD can be developed. For example, the RHD Australia 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: <ul style="list-style-type: none"> • 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. • The WHO Pocket Book of Hospital Care for Children includes information about management of RHD.

10. Programme evaluation

Things to consider

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

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.

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. 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.

Table 15: Approaches to programme evaluation

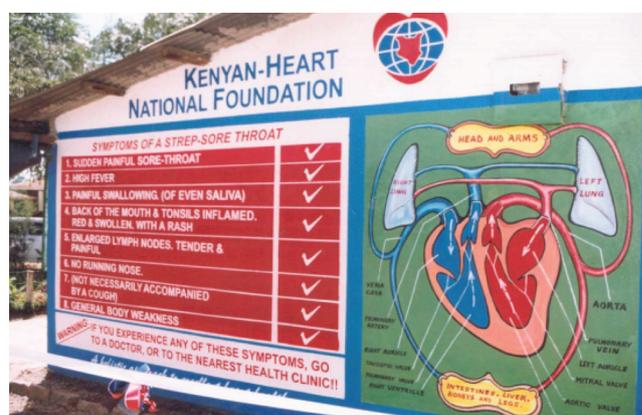
	Advantages	Disadvantages
<p>Narrative review Evaluation of RHD control programmes has been in narrative form, outlining the project and describing outcomes. Often these reports are required by donor agencies or other funding groups.</p>	<ul style="list-style-type: none"> • Can provide a compelling story of the programme and its impact. 	<ul style="list-style-type: none"> • Usually free text which can make it difficult to compare parameters and outcomes over time.
<p>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”.</p>	<ul style="list-style-type: none"> • Indicators comparable across time so improvement can be measured. • Actionable areas for improvement can be identified. 	<ul style="list-style-type: none"> • Indicators often do not exist and need to be developed for the audit. Can be time consuming to conduct.
<p>Continuous quality improvement (CQI) CQI is an action research process which shares similarities with the clinical audit cycle though it tends to be more comprehensive and designed to be an ongoing project. 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.</p>	<ul style="list-style-type: none"> • Can engage all members of the RHD control programme. • Offers a framework for ongoing review and improvement in service delivery. 	<ul style="list-style-type: none"> • Can take time and effort for staff to engage with the CQI process.
<p>Contracted or external evaluation External evaluation can provide important and independent assessment of how your programme functions. 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.</p>	<ul style="list-style-type: none"> • Provides an independent assessment of your programme. • May include detailed analysis of areas to strengthen. 	<ul style="list-style-type: none"> • May be expensive if privately contracted.

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.



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

11. Community education

Things to consider

- Who are your priority target groups?
- What do you want them to do?
- 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?

Table 16: Issues to consider in designing an awareness campaign

Participant involvement	Community members and organisations should be involved in the design and implementation of education programmes. This helps to ensure that valuable, culturally relevant messaging, best reflects the local needs.
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.
Needs and resources assessment	Existing community resources and infrastructure should be identified and evaluated to avoid duplication. New resources should be developed to meet the needs of your target audience, their existing knowledge and need for new information.
Comprehensive programme	Comprehensive RHD control programmes address community education at a variety of levels; including children, parents, teachers and community groups.
Integrated programme	Integrated education programmes provide consistent messages across all media and locations. In this way, health messages can be integrated into people’s lives.
Long-term change	A successful programme will establish a sturdy foundation that can serve as a platform for enduring change. Education should be established to promote sustainability, rather than occur as a “one-and-done” outreach effort.
Research and evaluation	Education campaigns should be evaluated during and after their implementation, to improve existing awareness programmes and inform the design of future campaigns. Pre and post intervention surveys can demonstrate acquisition of new knowledge.

Table 17: Media for communication

Pamphlets	Pamphlets are relatively low cost, easy to distribute and can be taken home by families for later reference. However, they will need to be adapted to low literacy settings.
Posters and billboards	Billboards, sidings, posters, wall paintings and public notices may be a valuable - and relatively low cost - opportunity to provide health promotion messages.
Films and video	Videos and films can be a powerful way of sharing information about RF and RHD. Producing videos can be time consuming and expensive so it is important to have a clear vision for how audio-visual material will used. Interviewing or involving policy makers in the film may be a way to engage their interest in RHD control.
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. Online community education modules are also emerging.
Radio and television	Radio and television messaging may be particularly useful for dispersed populations or in times of social instability and in areas of low literacy.
Celebrities	People who are famous or popular may be able to access adolescents who can be difficult to reach with traditional health promotion messages.
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. A Heart Club founded in Kerala, India hosts events for young people living with RHD.

12. Sore throat guidelines

Things to consider

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

In settings with endemic RF/RHD approximately two thirds of sore throats are caused by viral infections. One third of sore throats are caused by a bacterial infection, most commonly group A streptococci (GAS). Treatment with oral penicillin can reduce the attack rate of RF following GAS by about 70% and up to 80% with IM penicillin. A full 10 day course of appropriate antibiotic treatment started within nine days of sore throat symptoms can prevent almost all cases of RF. Distinguishing viral pharyngitis from bacterial pharyngitis is difficult. A wide range of clinical guidelines have been published by organisations in high resource settings. 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. Developing local guidelines for the diagnosis and treatment of sore throat is an important role for RF/RHD control programmes.

Treatment approaches

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

Antibiotic resistance

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

Table 17: Approaches to sore throat treatment

Treat all sore throats	In some countries the burden of RHD is high, access to health workers is limited, and laboratory services very poor. In these cases, programmes may choose to treat all children with signs and symptoms of significant sore throat. However, only a fraction of children receiving antibiotics will have a GAS bacterial sore throat which will benefit from treatment. In a ‘treat all’ model the majority of children won’t benefit from antibiotics, risking unwarranted adverse drug reactions and potentially contributing to antibiotic resistance.
Clinical scoring tool + treat	A range of clinical scoring tools have been developed to try and distinguish GAS pharyngitis from viral pharyngitis. 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.
Test sore throats for GAS and treat positives	In a test-and-treat model, patients with a sore throat have antibiotic therapy guided by biologic tests to distinguish GAS pharyngitis from viral pharyngitis. These tests are may include rapid antigen tests and/or throat culture (see Chapter 4). This approach supports rational use of antibiotics but may miss some cases of GAS via false negative antigen tests or because treatment is delayed whilst awaiting throat culture results. Programme costs in a test and treat model may be higher and may not be feasible in low resource settings. Swabbing techniques, sample storage and transfer are all important considerations if throat swabs are going to be part of your RHD control programme.

13. Provision of primary prophylaxis

Things to consider

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

Table 18 Challenges in delivering primary prophylaxis

<p>Patients and families</p>	<p>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. Providing education and information about the importance of sore throat treatment is vital for effective delivery of primary prophylaxis.</p> <p>Even when people want to seek care there may be barriers which prevent access, including</p> <ul style="list-style-type: none"> • 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.
<p>Individual health care providers</p>	<p>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. Strategies for improving use of guidelines may include:</p> <ul style="list-style-type: none"> • 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.
<p>The health care system</p>	<p>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. 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.</p>

“Two aspects are fundamental to any control programme. Firstly, the people in a community at risk must be aware of the problem, particularly those at high risk who require prophylaxis against recurrences. Secondly, the facilities for preventing the disease at primary and secondary levels must be easily accessible to those who need them.”

Edgerton et al, Soweto, South Africa, 1982.²³⁸

The role of chemists and pharmacists

In many parts of the world private pharmacies are the main source of medication and health advice. 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. Including and educating private providers in your RHD control programme may be one approach to improve delivery of primary prophylaxis.

14. Active case finding: sore throat clinics

Things to consider

- Are other school based health services delivered in your area?
- Could sore throat management be included in the role of school nurses or health workers?
- How would families and children consent for inclusion?
- Would there be a way to include 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. A small number of historic sore throat clinics were also run in the United States. 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).
- 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.
- Logistic challenges may be prohibitive in low and middle income country setting.

The challenge of low pharyngitis reports

The relationship between RF and preceding sore throats is complicated. Studies from New Zealand, Australia and Pakistan indicate that a significant proportion of children present with RF without any recollection of pharyngitis. Poor correlation between sore throat and RF may represent recall bias, asymptomatic infection or GAS infection from a skin source. 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.

15. Vaccine development

Things to consider

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

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. Progress towards a safe, effective, affordable and practical GAS vaccine has accelerated in recent years.

Prioritizing, developing and implementing vaccine programmes is a hugely complex international undertaking. Similarly, basic science development of vaccines is highly specialised, expensive and technically complex. 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. In light of these experiences WHO developed Vaccine Introduction Guidelines to help countries make decisions about new vaccines. This model has been expanded and adapted to explore preliminary work for a malaria vaccine, and a country planning for emerging health interventions. A framework for preparatory GAS vaccine engagement is needed. In the interim key topics adapted from malaria are outlined in table 19.

“Although the burden of disease was clearly necessary for adoption decisions, it was not generally sufficient; political prioritization was also very influential”

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

Table 19: Determinants of new vaccine adoption in lower-middle income countries

Signal vaccine demand	Identifying and communicating vaccine needs in low resource settings is critical for securing interest, funding and 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.
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.
Existing intervention data	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.

Secondary Prevention

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 enrollment into a register-based programme for antibiotic administrations. Diagnosis and registration is a fragile process. Worldwide, many people are thought to be undiagnosed; because symptoms are mild, or 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 aid facilitate follow up and regular antibiotic administration. In places with an RHD register it may be difficult to enroll, 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 antibiotics 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 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 – 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

16. The RF and RHD register

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?

A disease register is a list of people who have been diagnosed with, or are suspected of having a disease.

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. These registers helped provide newly developed regimes for antibiotic secondary prophylaxis and contributed to the declining burden of RHD in the USA. By the 1970s the WHO had adopted a register based approach and registers remain a critical part of RHD control worldwide.

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.

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.

Table 20: Benefits of RF/RHD registers

	For people living with RF/RHD	For health systems
	<p>Improves delivery of consistent, disease altering, secondary prophylaxis through:</p> <ul style="list-style-type: none"> • Recall systems • Standardised care delivery 	<ul style="list-style-type: none"> • 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

Table 21: Register location

	Advantages	Disadvantages
Local	<ul style="list-style-type: none"> • Useful for following up individual patient • Can be easily updated and accessed by local clinical staff 	<ul style="list-style-type: none"> • Can be difficult to transmit information to a central register, limited value as an epidemiologic tool
Central	<ul style="list-style-type: none"> • Provides a population overview of the burden of disease • Valuable for mobile populations or people who move frequently 	<ul style="list-style-type: none"> • Requires systems to provide information back to the health system level • May not be clinically relevant for primary care provision

Table 22: Register format

	Advantages	Disadvantages
Electronic	<ul style="list-style-type: none"> • Reports and data easily extracted • Supports integrated care if combined with electronic health records 	<ul style="list-style-type: none"> • May be more expensive to establish and maintain • Staff training may be required • Data may be lost
Paper	<ul style="list-style-type: none"> • Little training required 	<ul style="list-style-type: none"> • Data is difficult to extract and analyse • Data may be lost or misplaced

Table 23: Potential states for RHD registers

Additions		Status Changes	
After an episode of RF		Prophylaxis ceased or completed	
<p>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.</p> <p>If possible it is simplest to encourage clinicians to contact the register about <u>all</u> 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.</p>		<p>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.</p> <p>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.</p> <p>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.</p>	
After first presentation of symptomatic RHD		Inactive	
<p>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.</p>		<p>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’.</p>	
Following screening		Death	
<p>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.</p>		<p>Recording deaths of individuals on the RHD register is important in order to:</p> <ul style="list-style-type: none"> • 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. <p>In some places it may be possible to access hospital death records to identify people with RHD who have died. 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.</p>	
Transfer in from another programme		Transfer out to another programme	
<p>In some countries people living with RHD are a mobile population who move frequently for work, health care, family or traditional responsibilities. 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.</p> <p>Other sources of information for the register may include: notifications, hospital discharge records, clinical letters, echocardiogram reports or professional correspondence. These records may also provide valuable information about the clinical status of people already on the register.</p>		<p>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:</p> <ul style="list-style-type: none"> • 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. <p>Consent to share clinical information with other programmes should be obtained.</p>	

17. BPG and other antibiotic supply

Things to consider

- Is BPG on your national essential medicines list or formulary?
- Do you have stock outs or shortages of BPG?
- Do your patients have to pay for BPG? Is cost a barrier to adherence?
- Do you have a system for ensuring all people are asked about a history of penicillin allergy?
- Do you have guidelines for management of anaphylaxis?
- Do you have access to adrenaline and other treatments for anaphylaxis?
- Do you have mechanisms for monitoring or reporting adverse drug reactions?

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. 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.

Safety and anaphylaxis

Patients and health care providers are understandably concerned about the risk of anaphylaxis from BPG injections.

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.

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.

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.

Supply

Stock outs and shortages of BPG have occurred in high and low resource settings. In the event of a shortage or stock out of BPG the World Heart Federation recommends:

- 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.

18. Provision of secondary prophylaxis

Things to consider

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

Once your programme has a register of people living with RHD and has established a reliable supply of antibiotics, delivery of secondary prophylaxis can begin. Adherence to the local schedule of secondary prophylaxis injection is

poor worldwide. Adherence is consistently less than 50% of scheduled injections delivered in a range of settings.

Table 25: Contributors to poor adherence

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. Cultural beliefs, including the role of traditional medicines 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.
Cost	In some places there is a direct financial cost to receive BPG injections. In other places people must take time off work or school to travel for injections causing indirect costs in time and travel.

Table 26: Strategies to improve adherence

Identify regular staff to deliver injections	A single dedicated health care worker responsible for administering BPG and ensuring adherence seems to improve uptake. 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. In some countries cards are used for monitoring health delivery and outcomes, including immunisation cards and 'under 5' cards.
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.
Memory cues	In 2006, the Central Australia RHD control programme launched a 'full moon strategy' 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. Other opportunities may include provision of calendars or the development of a smartphone app.
Text messaging and phone calls	Text messages (SMS or texting) or phone calls can be used to remind people that secondary prophylaxis injections are due.
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. 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.
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.

19. Priority based follow up (clinical review)

Things to consider

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

Developing an RHD register helps to improve delivery of secondary prophylaxis. 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.

Table 27: Considerations for development of a priority based follow up system

Benefits of a priority system	Precautions of a priority system
<ul style="list-style-type: none"> • 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. 	<ul style="list-style-type: none"> • 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.

Table 28: Steps to developing a priority based system

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. Providing a standardised case management approach has been valuable for other conditions endemic in low resource settings.
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.

20. Active case finding (echocardiography screening)

Things to consider

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

An introduction to screening

Health screening programmes are designed to 'discover those among the apparently well who are in fact suffering from the disease'. 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.

The role of auscultation screening

Screening for RHD by auscultation alone is no longer appropriate where echocardiography is available.

Table 29: Rationale for echocardiography screening

To establish burden of disease (populations)	To influence clinical outcomes (individuals)
<ul style="list-style-type: none"> • Provides the best quality standard burden of disease data needed for planning and delivering programmes • A known denominator and standard diagnostic criteria are needed to be able to interpret the results of the screening project. • Although screening is being conducted to gather population information, individuals will also be affected. Systems to manage children diagnosed with RHD during screening will be needed. 	<ul style="list-style-type: none"> • The clinical significance of borderline asymptomatic heart valve abnormalities on echocardiography is unclear. • Echocardiography screening for clinical benefit is not currently recommended.

Table 30: Considerations for echocardiography screening

Echo criteria for diagnosis of RHD	The WHF Echo criteria are the universal standard for diagnosis of RHD in young people less than 20 years. For screening a simplified criteria that can be utilised by technicians with basic levels of training in echo may be used as an initial test.
Who should be screened	Age is an important determinant of RHD prevalence: older adolescents will have more detectable valve lesions than younger children. However, valve disease is likely to be more severe and so participants will receive less benefit from early detection. Children and adolescents who attend school are often the most accessible for screening. However, children who do not attend school are also at risk of RHD and approaches to identifying them will need to be considered.
Expert review of images	Large volumes can be time consuming and cause delays in interpretation.
Privacy	If there is a need to disrobe for the echo consider the need change rooms and gowns.
Consent	What are the local practices regarding the need for the provision of enough information for the person, parent or guardian to provide informed consent?
Recording outcomes and communicating results	Local doctors, nurses and health workers should be provided with documented results for the patients regular file and patients and families need to be informed of the results of the test.
Follow up	The logistics of how a patient will be managed immediately and into the future if they are found to have RHD or another problem, such as congenital heart disease need to be planned in advance.

Tertiary interventions

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 for clinical care, support of health care staff and programme sustainability.

Comprehensive RHD control programmes offer an unprecedented opportunity for diagonal health system strengthening and integrative care. 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.

Table 31: 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.	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.	Mitral stenosis generally develops in more advanced RHD and is often caused by persistent or recurrent inflammation of the mitral valve.	
Aortic Valve Valve between the left ventricle and the aorta.	Aortic regurgitation (AR) occurs when the aortic valve does not close properly. AR generally causes left sided heart failure.	Narrowing and scarring of the aortic valve can cause obstruction to left ventricular outflow. RHD is a rare cause of aortic stenosis.	
Tricuspid Valve Valve between the right atrium and the right ventricle.	Rarely, RHD can cause isolated damage to the tricuspid valve, generally regurgitation.		
Pulmonary Valve Valve between the right ventricle and the pulmonary artery.	The pulmonary valve is very rarely damaged by RHD.		

21. Diagnosis and management of RF & RHD

Things to consider

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

Table 32: Potential health system roles for RF management

Primary Care	Secondary Hospital	Tertiary Hospital	Quaternary
Suspected case of RF identified <ul style="list-style-type: none"> • Referred for secondary evaluation • Register notified of suspected case 	Admission and specialist evaluation <ul style="list-style-type: none"> • Definitive diagnosis made • Register notified of diagnosis • Referred to tertiary center if evidence of heart failure 	Admission for advanced medical management <ul style="list-style-type: none"> • Clinical management of heart failure • Referral to surgical center if required 	Admission if acute surgery required

Table 33: Sample discharge considerations for RF

Treatment	<ul style="list-style-type: none"> • Give the patient the first dose of secondary prophylaxis • Provide a prescription for pain relief from arthralgia if still required
Education	<ul style="list-style-type: none"> • Broad education to include: explanation of RF and RHD, importance of secondary prophylaxis, symptoms that may represent a recurrence
Referrals*	<ul style="list-style-type: none"> • Notify the RHD register coordinator of a new person to receive secondary prophylaxis • 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 <p>*Seek consent where required or appropriate</p>

Medical care of people living with RHD

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. Medical admissions for RHD tend to be for the treatment of heart failure, with diuretics. Admissions may be prolonged over weeks and families will require support over this time, particularly if inpatient facilities are a long distance from home villages or usual residence.

Medical care for women of child bearing age with RHD should also include information about safe pregnancies, or avoiding pregnancy. Locally available family planning, with a low failure rate, should be offered.

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. 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 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. Treatment for pain, nausea, constipation and anxiety may also be required. 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. Your programme may also provide support for families affected by deaths from RHD.

22. Anticoagulation

Things to consider

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

Anticoagulants are medications which make blood less likely to clot (coagulate). Anticoagulation is indicated for the management of symptomatic RHD for some patients with arrhythmia (particularly atrial fibrillation) and heart failure. People who have had mechanical heart valve replacements depend on effective anticoagulation for survival. Delivered effectively, anticoagulation prevents thrombus (clot) formation which could cause a stroke or block a mechanical heart valve. 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 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. The dose of the warfarin needs to be changed 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 34 and 35.

Table 34: Models of INR testing

Laboratory INR testing	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. Results take time to process and communicate, sometimes arriving days after the initial blood tests.
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. These machines require occasional testing/calibration and ongoing supply of reagent cartridges. Recommended technical specifications of POCT machines are available online.
Little-or-no testing, fixed dose warfarin	Where INR testing is impossible, people are sometimes prescribed a fixed dose of warfarin for months at a time without INR testing. This regime produces inferior clinical outcomes but may be the only possible approach in some settings.

Table 35: 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. This allows primary care doctors to provide comprehensive care for a range of medical conditions, and maintain frequent contact with people needing close monitoring.
Anticoagulation clinics	Management of anticoagulation is managed by special clinics in some places. This provides a clear focus on safe and effective anticoagulation, staff skilled in dose titration and experience with INR fluctuations. The time in therapeutic INR range may be improved compared with anticoagulation from primary care.
Pharmacist led anticoagulation	Anticoagulation education, dosing and monitoring may be arranged through pharmacy services. Strong pharmacy engagement appears to improve time in therapeutic INR range and minimise adverse anticoagulation events in comparison to standard primary care. 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.
Patient led anticoagulation	Some highly health literate patients in well-resourced settings have their own point of care INR machines and adjust their own warfarin dose within pre-specified limits.

23. Triage & preoperative evaluation

Things to consider

- How does your programme manage the list of people waiting for surgery?
- Does your programme have a relationship with a regular surgical or interventional service?
- How do you communicate 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?

Table 36: 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. The procedure is technically more difficult than valve replacement, and particularly difficult in RHD compared with other causes of mitral valve damage.
Valve replacement	Heart valve replacement is an open heart surgical procedure. Surgeons remove the damaged heart valve and replace it with a mechanical prosthetic (metallic valve) or bioprosthetic valve (tissue valve). Bioprosthetic valve replacements cause fewer blood clot complications than metal valves but are more likely to wear out and require replacement. Mechanical valve replacement is associated with high risk of embolism and haemorrhagic complications but usually lasts for life.
Balloon valvotomy (valvuloplasty, 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. The closed approach reduces costs and complications compared with open surgical repair, providing a safe and effective option for low resource settings. However, a cardiac catheterisation laboratory is required to perform the procedure and few facilities exist in the areas of greatest need.

Table 37: Pre-operative considerations

For the health system	For individuals
<p>Triage of intervention candidates is critical in places where surgical or percutaneous interventions are possible to ensure that the most appropriate patients receive the limited resources.</p> <p>It may be necessary to consider:</p> <ul style="list-style-type: none"> • Capacity of individual patients to benefit from surgery. • Ability of the surgical team to undertake 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. • In some programs evidence of compliance with secondary prophylaxis is considered in the context of preoperative evaluation <p>Planning for interventional care:</p> <p>Include an approximate estimate of the number of people who may benefit from pre-operative assessment and system capacity to deliver interventions.</p> <p>Who should be referred and when:</p> <p>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.</p>	<p>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. Considerations for pre-operative optimisation may include:</p> <p>Echocardiography: Echocardiography data provides critical information regarding valve lesions, cardiac chamber size, left ventricular function and pulmonary artery pressure. and serial data will assist with determining the timing of surgery.</p> <p>Dental optimisation: Dental optimisation prior to surgery is particularly important to reduce the risk of subsequent bacterial endocarditis.</p> <p>Nutrition: A person with a good nutritional status before surgery will have improved post-surgery outcomes compared to a person who is under nourished.</p> <p>Pregnancy status: Female surgical candidates being evacuated or travelling for surgery should have their pregnancy status confirmed before departure.</p> <p>Infectious disease status: Evaluation for potential infectious diseases will vary by setting but may include testing for tuberculosis, HIV, hepatitis C and malaria.</p> <p>Routine preoperative bloods: Liver function tests, creatinine, glucose, electrolytes.</p>

24. Post intervention review, follow up & audit

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?

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. 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.

“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.

Table 38: Models of care following intervention

Primary Care	Secondary Hospital	Tertiary Hospital
<ul style="list-style-type: none"> • Monitoring of complications • Repeat prescriptions 	<ul style="list-style-type: none"> • Potential for ‘step down’ or convalescent care • Management of complications • INR monitoring 	<ul style="list-style-type: none"> • Discharge education • Anticoagulation initiated if required • Follow up appointments scheduled

← Documentation and care plan

Table 39: Post operative planning

1. Post-operative planning for the health system	In the same way that GAS treatment guidelines and secondary prophylaxis guidelines help to standardise care, a structured pathway of care around the time of a heart operation is also needed. The aim is to improve the quality and safety of care provided which meets the needs of individuals, families and your local capacity for service delivery. A default 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.
2. Post-operative planning for individuals	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. Communication with patients and families is essential to explain that surgery is not a definitive solution for RHD. Most post-operative patients will require lifelong anticoagulation, secondary prophylaxis and medical supervision of pregnancies. 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.
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. Where possible, cases should be presented at regular institutional meetings and be open to peer review. 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. 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.

25. Provision of interventional services

Things to consider

- Does your programme interact with any interventional service providers?
- Are these interactions be mutually beneficial?
- Is the current model of interventional service provision sustainable?

A minority of countries with a high burden of RHD have local access to surgical interventions for the disease. A variety of approaches for delivery of surgical services have evolved to address this unmet need. These models are summarised in table 40.

Surgeons as health systems advocates

Surgical teams have a rare and valuable opportunity to advocate for addressing the underlying causes of RHD. Surgical teams should be encouraged and supported to discuss the need for prophylaxis, pre-operative and post-operative care.

Table 40: Models of surgical care for RHD

		Location of the procedure	
		Local or national	International
Usual location of the surgical team	Local or national	<p>National /regional centers of excellence</p> <p>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.</p> <p>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.</p>	<p>International surgical training</p> <p>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.</p> <p>International placements to develop surgical skills in high resource settings play an important role in transferring skills to new surgeons and teams. Challenges include may include supporting surgical staff from low resource settings to return home following international training and ensuring sufficient case volume to acquire surgical skill in RHD which is rare in high resource settings.</p>
	International	<p>Humanitarian surgical missions</p> <p>In some resource limited settings ‘fly in fly out’ teams are an important component local health services. Humanitarian cardiac service delivery is supported by a number of international organisations. While there are considerable benefits from international cardiac surgery missions there are also enormous challenges, and significant potential for harm.</p>	<p>Surgical evacuation</p> <p>In some countries patients must travel internationally for operations. This may be arranged formally through the health system or privately by individuals or families. In some settings specialists visit for triage and follow up, while operations and interventions occur remotely. This model may occur within countries or between countries. Private arrangements made by families are less likely to be subject to triage or 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.</p>

Annex A: Assessment

The TIPs approach outlined in this handbook emphasises the importance of addressing the component of comprehensive RHD control programmes. Identifying the activities in each domain makes it possible to strengthen or celebrate success in each chapter. To support this descriptive process an assessment tool has been developed to help define and describe how your programme address the chapters outlined in TIPs. The TIPs Assessment Tool provides an valuable opportunity to define and describe existing work when beginning, reviewing or revising RHD control programmes. Working through the assessment may help to identify areas which would otherwise be overlooked or under-addressed during programme delivery. 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/>

