



**WORLD HEART  
FEDERATION®**

**Diagnosis and Management of**

**Acute Rheumatic Fever  
Rheumatic Heart Disease**

This training curriculum has been developed to assist the reader to

1. Identify the causes of acute rheumatic fever (ARF) and rheumatic heart disease (RHD)
2. Identify individuals and groups who are most at risk
3. Recognise the signs and symptoms of ARF and RHD
4. Identify standard drugs, doses and regimens used to prevent recurrent ARF
5. Describe priority management of people with ARF and RHD
6. Evaluate and report on local RHD control strategies and rates of disease.

This document is intended for training health workers and others involved in the diagnosis and management of acute rheumatic fever and rheumatic heart disease.

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## 1 INTRODUCTION

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Acute Rheumatic Fever (ARF) and Rheumatic Heart Disease (RHD) is the most common cardiovascular disease in children and young adults and remain a major public health problem in developing countries. Recent research estimates that about 15.6 million people are affected worldwide, 2.4 million of who are children between 5 and 14 years old in developing countries. Almost half a million new cases are declared every year. ARF and RHD result in an estimated 350,000 deaths annually, and hundreds of thousands of survivors are left disabled without access to the expensive medical and surgical care that RHD requires.

ARF is a disease of poverty. It is particularly rampant in low-income, overcrowded communities with poor housing conditions, poor nutrition and inadequate health services.

An untreated Group A streptococcal infection can lead to ARF. ARF is an autoimmune condition which mainly affects the large joints (arthritis) and the heart (carditis). Repeated Group A streptococcal infections and recurrent ARF can lead to chronic heart valve damage (RHD) requiring expensive heart valve surgery. If damaged heart valves are not repaired or replaced by major open-heart surgery, the condition is often fatal.

RHD is preventable.

- The first episode of ARF can be prevented by treating Group A streptococcal infections with penicillin;
- If the first ARF episode is not prevented, recurrent episodes (which almost always lead to RHD) can be prevented with long-term penicillin delivered at regular intervals.

*Secondary prevention* refers to the delivery of regular penicillin to prevent further ARF and the development or worsening of RHD. Secondary prevention is best delivered as part of a programme that includes health education for parents, children, teachers and health care providers, routine screening for disease, availability of penicillin and maintenance of a disease register to manage known ARF and RHD cases. Importantly, secondary prevention programmes can be implemented through the existing health care services without major additional costs.

### 1.1 Definitions

#### 1.1.1 Group A beta-haemolytic *streptococci*

Humans are exposed to Group A streptococcus (GAS) bacteria through the environment. Infections of the throat and skin are common sites of infection. GAS infections of the throat have been positively linked to the onset of ARF.

GAS throat infections occur commonly in children throughout the world, with the peak ages between 5 and 15 years. The number of children affected in each region varies depending on environmental conditions, level of poverty and the quality and availability of health care.

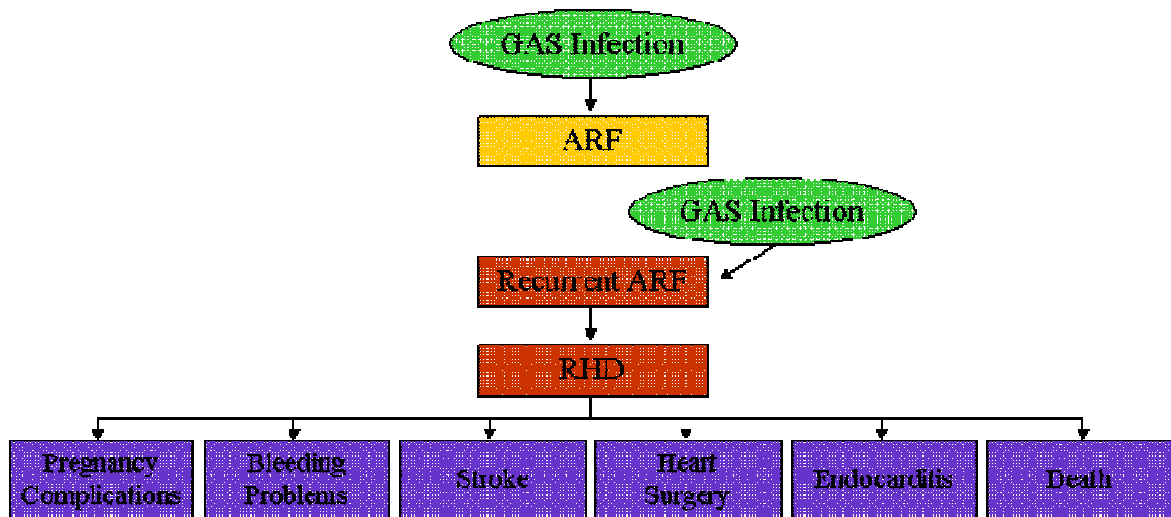
#### 1.1.2 Acute Rheumatic Fever

Acute rheumatic fever (ARF) is a delayed autoimmune response to an untreated GAS infection. ARF may involve the heart, joints, central nervous system and/or skin. Signs and symptoms may include any or all of the following: arthritis (of one or more joints), fever, carditis (inflammation of the heart), rash, Sydenham's chorea (uncontrolled movements), and subcutaneous nodules.

ARF develops about 2-3 weeks after the onset of a GAS infection. The illness usually lasts up to 3 months and resolves without treatment. With treatment the symptoms resolve within 1-2 weeks. ARF can occur repeatedly in people who continue to be exposed to high levels group A streptococci in their environment.

#### 1.1.3 Rheumatic Heart Disease

Rheumatic heart disease (RHD) presents as damage to the heart valves as the result of repeated attacks of ARF. The valves become stretched and scarred and do not move normally. The valves may not close properly which can allow blood to leak backwards, and/or the valves may not open properly which can cause blood flow to be blocked. If RHD is not diagnosed and managed early, it may result in heart failure and premature death.



**Diagram 1.** Progression of Disease.

#### 1.1.4 Disease Control

ARF or RHD can be diagnosed in individuals at any stage during disease progression. Early diagnosis of ARF and regular treatment can prevent the development of RHD.

ARF and RHD they can be prevented at population level by sustainable control strategies. For regions with high rates of disease the World Health Organisation (WHO) recommends a dedicated, register-based programme which focuses on identifying cases, delivering regular prophylaxis treatment, and education. Programmes have been established in many countries throughout the world. Success of RHD control programmes is determined by the level of Government commitment to funding and resources, capability and enthusiasm of staff involved, and response from the community.

This training package has been designed to provide an understanding of ARF and RHD control and assist in the development of sustainable and effective control programmes.

## 2 EPIDEMIOLOGY

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### 2.1 Risk Factors for ARF

Rheumatic heart disease is a disease of poverty. The following factors increase the risk of developing ARF:

- Overcrowding and poor standard of housing;
- Reduced access to health care;
- Living in a tropical climate.

ARF is most common in children between the ages of 5 and 15 years.

ARF is less common after the age of 35 years.

ARF is rare under 4 years and over 40 years of age.

*NOTE: ARF should be considered in all age groups in high risk populations, and not ruled out because of age.*

### 2.2 Changing pattern of RHD

Improved living conditions, a better standard of health care, and the introduction and use of antibiotics have decreased the prevalence of ARF and RHD in most developed countries over the past century. RHD is still common in developing countries, and among indigenous populations in developed countries.

### 2.3 Epidemiology of ARF and RHD

In 1994, it was estimated that 12 million individuals suffered from ARF and RHD worldwide<sup>[1]</sup> and at least 3 million had congestive heart failure (CHF) that required repeated hospital admissions<sup>[2]</sup>. A large proportion of the individuals required cardiac valve surgery within 5–10 years<sup>[1-3]</sup>. The death rate from RHD varied from 0.5 per 100,000 population in Denmark, to 8.2 per 100,000 population in China, and the estimated annual number of deaths from RHD for 2000 was 332,000 worldwide<sup>[4]</sup>. The death rate per 100,000 people varied from 1.8 in the WHO Region of the Americas, to 7.6 in WHO South-East Asia Region. Information from developing countries suggests that death due to ARF and RHD remains a problem, and that children and young adults still die from ARF<sup>[1-5]</sup>. A recent systematic review of 57 studies found the highest calculated prevalence in sub-Saharan Africa, the Pacific and Indigenous Australia and New Zealand. (Table 1) Reliable information on the incidence of ARF is scarce. In some countries, however, information from local ARF registers of schoolchildren provide useful information on trends.<sup>[4]</sup>

The annual incidence of ARF in developed countries began to decrease in the 20th century, with a marked decrease after the 1950s; it is now below 1.0 per 100,000<sup>[6]</sup>. The few studies conducted in developing countries report incidence rates ranging from 1.0 per 100,000 school-age children in Costa Rica<sup>[7]</sup>, up to 72.2 per 100,000 in French Polynesia, 100 per 100,000 in Sudan, and 150 per 100,000 in China. The incidence in Aboriginal children in the Top End of Australia's Northern Territory is 330 per 100,000.

The prevalence of RHD has also been estimated in surveys, mainly of school-age children. Survey results showed there is a wide variation between countries, ranging from 0.2 per 1000 schoolchildren in Havana, Cuba<sup>[8]</sup> to 2.2 cases per 1000 in Cambodia and 2.3 cases per 1000 in Mozambique<sup>[9]</sup>. The prevalence of ARF and RHD and the mortality rates vary widely between countries and between population groups in the same country, such as between Maoris and non-Maoris in New Zealand, Samoans and Chinese in Hawaii, and Aboriginals and non-Aboriginals in Northern Australia<sup>[1, 4, 10]</sup>.

Although it is known that death figures from hospitals often give biased information about the amount of diseases in the community, they are the only sources of information available in many developing countries. Based on this, RHD accounts for 12–65% of hospital admissions related to cardiovascular disease, and for 2.0–9.9% of all hospital discharges in some developing countries. There has been a marked decrease in the RHD-related death, incidence, prevalence, hospital admissions and severity of ARF and RHD in some places that have implemented prevention programmes, such as; Havana, Cuba; Costa Rica; Cairo, Egypt; and Martinique and Guadeloupe<sup>[4]</sup>.

### 2.4 Determinants of the disease burden of ARF and RHD

It is well known that socioeconomic and environmental factors play an indirect, but important, role in the prevalence and severity of ARF and RHD. Factors such as a shortage of resources for providing quality health care, inadequate expertise of health-care providers, and a low level of awareness of the disease in the community can all effect the expression of the disease in populations. Crowding adversely affects rheumatic fever incidence (Table 2).

Year	Area surveyed	Prevalence	Ages
2004 <sup>[11]</sup>	All developing countries	1.6 per 1,000	5-14 years
2007 <sup>[12]</sup>	Aboriginal Aust (Top End)	8.6 per 1,000	5-14 years
2007 <sup>[9]</sup>	Cambodia	2.2 per 1,000	6-17 years
2007 <sup>[9]</sup>	Mozambique	2.3 per 1,000	6-17 years
2008 <sup>[13]</sup>	Tonga	33.2 per 1,000	10-15 years
2008 <sup>i</sup>	Fiji	78 per 1,000	5-14 years

**Table 1.** International RHD Prevalence (Confirmed on echocardiogram)

Determinants	Effects	Impact on ARF & RHD burden
<p>Socioeconomic &amp; environmental factors</p> <ol style="list-style-type: none"> <li>Poverty</li> <li>Poor nutrition</li> <li>Overcrowding</li> <li>Poor standard of housing.</li> </ol>	<ol style="list-style-type: none"> <li>Rapid spread of Group A streptococcal strains</li> <li>Difficulties accessing health care</li> </ol>	<ol style="list-style-type: none"> <li>Higher incidence of acute strep pharyngitis and complications</li> <li>Higher incidence of ARF and recurrent ARF.</li> </ol>
<p>Health system related factors:</p> <ol style="list-style-type: none"> <li>Shortages of resources for health care</li> <li>Low level of knowledge of disease among health-care providers</li> <li>Low-level of awareness of the disease in the community.</li> </ol>	<ol style="list-style-type: none"> <li>Inadequate diagnosis and treatment of strep pharyngitis</li> <li>Misdiagnosis or late diagnosis of ARF</li> <li>Inadequate secondary prophylaxis delivery.</li> </ol>	<ol style="list-style-type: none"> <li>Higher incidence of ARF and recurrent ARF</li> <li>Missed first ARF episode.</li> <li>Inadequate secondary prophylaxis delivery</li> <li>Higher rates of recurrent ARF with more frequent and severe heart valve involvement.</li> <li>Higher rates of repeated hospital admissions and expensive heart valve surgery.</li> </ol>

**Table 2.** Direct and indirect results of environmental and health-system determinants on ARF/RHD<sup>[4]</sup>

<sup>i</sup> RHD School Screening, unpublished data

### 3 DIAGNOSIS AND MANAGEMENT OF ACUTE RHEUMATIC FEVER

#### 3.1 Diagnosis of Acute Rheumatic Fever (ARF)

##### 3.1.1 Revised Jones Criteria <sup>(4, 12)</sup>

Acute rheumatic fever was first described as an illness in *The Lancet* journal in 1889. The Jones Criteria were developed in 1944 as a set of clinical guidelines to help clinicians make a diagnosis of ARF. The Jones Criteria have been modified and altered a number of times, most recently by the World Health Organisation in 2003.

ARF diagnosis can be missed or delayed because

- A combination of signs and symptoms are required to confirm the diagnosis,
- ARF may be confused with other diseases which have similar sign and symptoms;
- People with ARF symptoms do not always present to the health system,
- Health workers may have difficulty recognising the signs and symptoms of ARF

The Jones Criteria include *Major* criteria, *Minor* manifestations, and evidence of a preceding GAS infection.

- *Major* manifestations are signs and symptoms more often associated with ARF
- *Minor* manifestations are signs and symptoms that can be included to help support the diagnosis.

Major manifestations:	Minor manifestations:	GAS Infection:
Carditis	Fever	GAS on throat swab (culture)
Arthritis	Arthralgia	Raised Anti-streptolysin O titre (ASOT)
Sydenham's Chorea	Prolonged P-R interval on ECG	Raised Anti-deoxyribonuclease B (Anti-DNase B)
Erythema marginatum	Raised ESR or CRP	
Subcutaneous nodules		

**Table 3.** Criteria for ARF diagnosis

According to the World Health Organisation <sup>(4)</sup>:

- The first episode of ARF can be confirmed if
  - 2 MAJOR, **or** 1 MAJOR and 2 MINOR manifestations are present **plus** there is evidence of preceding Group A streptococcal infection.
- Recurrent ARF (with no RHD) can be confirmed if
  - 2 MAJOR, **or** 1 MAJOR and 2 MINOR manifestations are present **plus** there is evidence of preceding Group A streptococcal infection.
- Recurrent ARF (with existing RHD) can be confirmed if
  - 2 MINOR manifestations are present **plus** there is evidence of preceding Group A streptococcal infection.

The World Health Organisation guideline is the international standard for diagnosis based on Revised Jones Criteria; however different regions have developed slightly modified guidelines to assist clinicians with local variations in ARF presentation <sup>(6)</sup> For example the following variations are included for consideration of ARF in some countries:

- the involvement of only one joint (mono-arthritis);
- poly-arthralgia in children who are at high risk of ARF (instead of poly-arthritis);
- sub-clinical carditis (evidence of rheumatic valve disease on echocardiogram)

**CHECK LOCAL GUIDELINES (if available):**  
**Criteria for acute rheumatic fever diagnosis may vary**

##### 3.1.2 Clinical Presentation <sup>(4)</sup>

The appearance of ARF varies among individuals and between populations. Occasionally, a sore throat may have resolved 1-2 weeks before other symptoms. People with ARF may present with the following:

- Arthritis (the most common symptom, in up to 75% of first episodes)
  - Pain, redness and swelling in the joints (commonly the ankles, knees, wrists, elbows, less commonly the small joints of the hands, feet and neck)
  - Often the first complaint
  - Usually 'migratory'- disappearing in one joint as it begins in another.
- Fever
- Carditis (inflammation of the heart)
  - Commonly presents as a heart murmur
  - Chest pain and/or difficulty breathing may be present in more severe cases
- (Sydenham's) Chorea
  - Twitchy, jerking movements and muscle weakness (most obvious in the face, hands and feet)
  - May occur on both sides or only one side of body
  - More common in teenagers and females (rare after age 20)
  - May be associated with irritability and or depression
  - May begin up to 3-4 months after the streptococcal infection, and often occurs without other symptoms
  - Usually resolves within 6 weeks (rarely lasts 6 months or more)
  - May recur in females during pregnancy

Less commonly, people with ARF present with the following:

- Subcutaneous nodules
  - Painless lumps on the outside surfaces of elbows, wrists, knees, ankles in groups of 3-4 (up to 12)
  - The skin is not red or inflamed
  - Last 1-2 weeks (rarely more than 1 month)
  - Nodules are more common when Carditis is also present
- Erythema marginatum
  - Painless, flat pink patches on the skin that spread outward in a circular pattern
  - Usually occurs early, may last months, rarely lasts years
  - Usually on the back or front of body, almost never on the face
  - Hard to see in dark-skinned people
- Cough and Abdominal pain

### 3.1.3 GAS infection

Evidence of a GAS infection is required to confirm a case of ARF with the above signs and symptoms.

- Note: Group A beta-haemolytic *streptococci* may not be found on a throat swab since the infection may be resolved at the time of onset of ARF symptoms.
- Serum ASOT –reaches a peak level around 3-6 weeks after infection and starts to fall at 6-8 weeks
- Serum Anti DNase B – reaches a peak level up to 6-8 weeks after infection and starts to fall at around 3 months after the infection

**CHECK LOCAL GUIDELINES (if available):**  
**Normal antibody titre ranges vary with age and geography**

## 3.2 Management of Acute Rheumatic Fever <sup>(12)</sup>

Persons with symptoms of ARF should be hospitalized to ensure accurate diagnosis, and to receive clinical care and education about preventing further episodes of ARF. The diagnosis should include an initial echocardiogram (if available) used to help identify and measure heart valve damage. Long-term preventative management should be organized before discharge.

### 3.2.1 Treatment of the acute illness

All cases of ARF should receive

- A single injection of Benzathine penicillin G, **or**
- Oral Penicillin for 10 days (Erythromycin if penicillin allergy) (refer to 4.3 *Standard Dose and Frequency*)

### 3.2.2 Relief of symptoms

Arthritis and fever

- Aspirin (Aspirin may hide symptoms of polyarthritis and fever. Paracetamol can be used until the diagnosis is confirmed)

#### Chorea

- Most mild-moderate cases do not need medication
- Provide calm and supportive environment (prevent accidental self-harm)
- Carbamazepine or Valproic acid can be given for severe cases.

#### Carditis

- Bed rest if in cardiac failure
- Anti-failure medication (e.g. Diuretics, ACEi, Digoxin)
- Anti-coagulation medication if atrial fibrillation is present

#### 3.2.3 Management plan when the acute episode is controlled

- Administer the first dose of secondary prophylaxis (refer to 4.3 *Standard Dose and Frequency*)
- Register the individual with the local health authority or RHD Programme;
- Provide disease education for the person with ARF and the family
  - Understanding of ARF and RHD and risks of ARF recurrence
  - Importance of regular secondary prophylaxis and medical review
  - Recognising own signs and symptoms of ARF and RHD
  - Risks associated with future RHD (e.g. pregnancy, surgery and high level of aftercare)
  - Importance of dental health
- Include an ARF diagnosis alert on computer systems and/or medical files (if applicable);
- Refer to local Medical Officer / health facility for ongoing management;
- Organise future specialist reviews (if required);
- Arrange dental review (and provide advice about endocarditis prevention);
- Influenza vaccination (if available).

#### 3.2.4 Long-term Management

Regular secondary prophylaxis (refer to 5.5 *Table 6 Recommended Secondary Prophylaxis Regimen*)

Regular medical review

Regular dental review

Echocardiogram (if available) following each episode of ARF, and routine echocardiogram:

- every 2 years for children (sooner if there is evidence of cardiac symptoms)
- every 5 years for adults (sooner if there is evidence of cardiac symptoms)

**CHECK LOCAL GUIDELINES:  
Management plans for acute rheumatic fever may vary**

#### 3.3 Management of Unconfirmed Rheumatic Fever

If an ARF diagnosis is considered highly probable (but not confirmed using the Jones Criteria) management should be commenced as above (section 3.2) in case it is an uncertain, but true diagnosis (refer to Table 3). Treatment and management should commence as for confirmed ARF with a Medical Officer review after one month (repeat echocardiogram if available) to detect the appearance of any heart valve lesions.

If there is evidence of rheumatic valve disease clinically or on echocardiogram, the diagnosis should be confirmed and a secondary prophylaxis regimen commenced.

## 4 SECONDARY PROPHYLAXIS

### 4.1 Introduction

Secondary prophylaxis is the terms used to describe regular delivery of antibiotics to prevent recurrence of GAS infection and subsequent development of ARF. Secondary prophylaxis is recommended for all people who have a history of ARF or RHD. The most effective method of secondary prophylaxis is Benzathine penicillin G given by intramuscular injection every 3 or 4 weeks <sup>(4)</sup>. Oral Penicillin may be used; however adhering to twice-daily tablets over many years is difficult. Oral Erythromycin is used if there is an allergy to Penicillin.

**Antibiotics need to be present in the body at all times  
to effectively prevent GAS infections which can result in recurrent ARF**

Regular secondary prophylaxis

- Prevents the occurrence of GAS infections which can lead to recurrent ARF
- Reduces the severity of RHD (and can result in cure of RHD after many years)
- Helps prevent death from severe RHD.

### 4.2 Indications for Use

Secondary prophylaxis is indicated for people who have

- ARF confirmed by the Jones Criteria
- RHD confirmed on echocardiogram
- ARF or RHD not confirmed, but highly suspected.

### 4.3 Standard Dose and Frequency <sup>(4)</sup>

#### 4.3.1 Benzathine Penicillin G

Injected Benzathine penicillin G is the most effective method of secondary prophylaxis. It has been shown to be effective against GAS infections in most people for 3 to 4 weeks. Benzathine penicillin G is usually given by deep intramuscular injection every 4 weeks, although it may be given every 3 weeks if ARF recurs despite regular 4-weekly injections or in high prevalence areas as indicated by local guidelines. The standard dose is

- **1,200,000 units for ALL people ≥30kg**
- **600,000 units for children <30kg**

#### 4.3.2 Penicillin V

Oral penicillin can be used if Benzathine penicillin injections are not tolerated or if injections are contraindicated. The standard dose is **250mg oral, twice-daily for ALL people.**

#### 4.3.3 Erythromycin

Erythromycin is given if there is a proven allergy to Penicillin. The standard dose is **250mg oral, twice-daily for ALL people.**

Drug	Route	Used for secondary prophylaxis of ARF
Benzathine penicillin G	Intramuscular	<b>YES</b>
Phenoxymethylpenicillin V	Oral	<b>YES</b>
<i>Benzyl penicillin</i>	<i>Intramuscular / intravenous</i>	<i>NO</i>
<i>Procaine penicillin</i>	<i>Intramuscular</i>	<i>NO</i>

**Table 4.** Penicillin used for secondary prophylaxis of ARF

### 4.4 Contraindications and Precautions

Allergic reactions to Benzathine penicillin G injections are rare and fatal reactions are very rare. The risk of allergic reaction does not appear to increase with long-term use. People do not become 'immune' to penicillin over a long period of time.

Benzathine penicillin G and Penicillin V should not be given to persons with a proven, serious penicillin allergy (refer to 5.8 *Drug Allergy and Anaphylaxis*). Clinicians should ask about previous drug allergies before commencing secondary prophylaxis. Any suspected Penicillin allergy should be confirmed before Erythromycin is used.

Penicillin causes no risk to the foetus and should be continued during pregnancy. Erythromycin is also safe and can be continued during pregnancy to prevent ARF. An ARF illness during pregnancy may cause serious risk to the mother.

Benzathine penicillin injections should be continued during anticoagulant therapy (Warfarin) unless there is a major intramuscular bleed following injection despite normal INR.

#### 4.5 Factors Influencing Secondary Prophylaxis Duration <sup>(4)</sup>

Every person who requires secondary prophylaxis should be evaluated independently as there are a number of additional factors that can help determine the length of time required for treatment. The following points should be considered carefully when planning the duration of secondary prophylaxis.

1. **Age.** ARF recurrence is less common after age 25 and rare after age 40, therefore younger people may need treatment for longer.
2. **Severity of RHD.** An additional ARF illness could be life-threatening for people with moderate or severe RHD and following valve surgery, therefore people with more complex disease may need to remain on treatment.
3. **Carditis during initial ARF.** Early heart damage increases the risk of further damage with recurrent ARF.
4. **Length of time since last ARF.** ARF recurrence is less common more than 5yrs since last episode.
5. **Medication delivery.** Regular prophylaxis in the first few years after the initial ARF may provide greater protection from recurrences than irregular prophylaxis for many years.
6. **Disease progression.** Evidence of worsening RHD at any stage may require extended prophylaxis.

Disease Classification	Duration of Secondary Prophylaxis
<b>ARF</b> (No proven carditis)	1. Minimum of 5 years after last ARF, or 2. Until age 18 years ( <i>whichever is longer</i> )
<b>Mild-moderate RHD</b> (or healed carditis)	1. Minimum 10 years after last ARF, or 2. Until age 25 years ( <i>whichever is longer</i> )
<b>Severe RHD</b> and following <b>Cardiac Surgery</b> for RHD	Continue medication for life

**Table 5.** Recommended Secondary Prophylaxis Regimens <sup>(1)</sup>

**CHECK LOCAL GUIDELINES:  
Secondary prophylaxis doses, frequency and/or regimens may vary**

#### 4.6 Ceasing Secondary Prophylaxis

The expected date to cease secondary prophylaxis should be recorded. Each person should be carefully reviewed to determine any ongoing risk of ARF recurrence and the level of heart valve damage before ceasing secondary prophylaxis. ***Ceasing secondary prophylaxis should only be done by a Specialist Medical Officer.*** Assessment prior to ceasing secondary prophylaxis should include

- Estimated time since last ARF illness (i.e. more than 5 years since last ARF)
- Specialist clinical review by a Medical Specialist or Heart Specialist
- Echocardiogram to establish presence and severity of RHD (if available).

#### 4.7 Benzathine Penicillin Injection Delivery

Benzathine penicillin G is most effectively given as deep intramuscular injection, into the upper outer quadrant of the buttock or the antero-lateral thigh.

##### 4.7.1 Assessment and Preparation

- Confirm the person's identity
- Review known drug allergies
- Discuss and record any ARF or RHD symptoms since last injection (refer to Medical Specialist if necessary)
- Obtain consent for injection

#### 4.7.2 Injection Procedure

A new, disposable syringe and needle must be used for each injection.

Benzathine penicillin G should be given immediately following preparation:

1. Check medication name and expiry date
2. Use a size 23 gauge needle for injection
3. Dispose of used needles and syringes in a puncture-proof container.

Prepare Benzathine penicillin G solution as directed by the product information:

- a. Administer 1,200,000 units for all persons  $\geq 30$ kg
- b. Administer 600,000 units for children  $< 30$ kg

**Pre-loaded syringes should be used only once.  
Squirt out unwanted solution before administering the required dose.**

#### 4.7.3 Documentation

- The following should be recorded in the Benzathine Penicillin injection book and/or medical record
  - Drug name, dose and batch number
  - Date injection given and date next due
  - Signature (of person giving the injection)
- Record the next date due on the person's clinic reminder card (if applicable)
- Provide the above information to the local RHD register/programme (if applicable).

#### 4.7.4 Pain Reduction

The following strategies may help to reduce pain<sup>ii</sup> following injection:

- Warm cold syringe to room temperature between hands (if applicable)
- Apply gentle pressure to the injection site for 10 seconds with the finger or thumb before injection
- Ensure that skin swabbed with alcohol is dry before injecting
- Deliver the injection slowly (preferably over 2 or 3 minutes)
- Use distraction to focus attention away from the injection
- Encourage movement (e.g. walking) following injection

### 4.8 Drug Allergy and Anaphylaxis

#### 4.8.1 Symptoms

Allergy to penicillin normally involves skin rash, and itchy eyes.

Rarely, allergy can cause an **anaphylactic reaction**, which can be fatal. Anaphylaxis occurs suddenly (within one hour) and symptoms can include wheezing, hives, itching, swelling of the face and lips, difficulty breathing, vomiting, a severe drop in blood pressure, loss of consciousness and cardiac arrest (heart stops beating). Vasovagal reactions (fainting) do not represent true anaphylaxis.

The risk of a serious reaction is reduced in children under the age of 12 years, and the duration of prophylaxis does not appear to increase the risk of an allergic reaction. The World Health Organisation <sup>(1)</sup> reports that allergic reactions to monthly Benzathine penicillin injections are rare:

Allergic reaction - 3.2%  
Anaphylaxis - 0.2%

#### 4.8.2 Treatment

An emergency kit for treating anaphylaxis should be available wherever Benzathine penicillin G is administered. All health workers administering secondary prophylaxis should also be trained in treating anaphylaxis.

Mild symptoms of allergic reaction can be treated with antihistamines.

Anaphylaxis should be treated with Adrenaline.

**CHECK LOCAL GUIDELINES:  
Adrenaline dose and administration may vary.**

<sup>ii</sup> Lignocaine 1ml 1% added to the syringe following preparation has been used successfully in some areas, however more research is required before it is a recommended routine practice

## 5 DIAGNOSIS AND MANAGEMENT OF RHD

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### 5.1 Diagnosis of RHD <sup>(4, 12)</sup>

#### 5.1.1 Introduction

Rheumatic heart disease is the result of damage to the heart valves which occur after repeated episodes of ARF. Early diagnosis of RHD is very important so that secondary prophylaxis can be started as soon as possible to help prevent the progression of the valve disease (see *Chapter 4 Secondary Prophylaxis*). Echocardiography is essential to confirm the diagnosis, and monitor the heart valves to detect any progression of disease.

The mitral valve is affected in over 90% of cases of rheumatic heart disease. The next most commonly affected valve is the aortic valve; usually disease of the aortic valve is associated with disease of the mitral valve. The tricuspid and pulmonary valves are rarely directly affected but tricuspid regurgitation may occur in advanced mitral valve disease.

Mitral regurgitation is the most common heart valve lesion in RHD – as an isolated lesion it is found most commonly in children and young adults. Mitral stenosis represents longer term chronic changes to the mitral valve – it is therefore more commonly seen in adults. A common complication of mitral stenosis is atrial fibrillation. Aortic regurgitation is not uncommon but aortic stenosis is almost never seen as an isolated lesion.

#### 5.1.2 Symptoms

The symptoms of RHD depend on the valve lesion and its severity. Symptoms of RHD may not show for many years until valve disease becomes severe.

In general terms, initial symptoms of RHD are the symptoms of early heart failure:

- Breathlessness on exertion
- Feeling tired
- General weakness

As heart failure progresses, other symptoms may develop including:

- Orthopnea (breathlessness on lying down)
- Paroxysmal nocturnal dyspnoea (waking at night with shortness of breath)
- Peripheral oedema

Palpitations may occur if atrial fibrillation is present (particularly in mitral stenosis). This arrhythmia is associated with increased thromboembolic risk including strokes. People with aortic valve disease may experience angina and syncope in addition to heart failure symptoms.

#### 5.1.3 Physical Examination

Clinical assessment should be conducted carefully because early detection of RHD can result in a better outcome. Careful auscultation should be undertaken and suspicious murmurs referred for assessment by a medical specialist with echocardiography (if available). Clinical examination should include assessment of severity and complications including signs of heart failure, the presence of atrial fibrillation and any new murmurs.

In **mitral regurgitation** the characteristic murmur is a pansystolic murmur heard loudest at the apex and radiating laterally to the axilla.

In **mitral stenosis** the characteristic murmur is a low-pitched, diastolic rumble heard best at the apex with the bell of the stethoscope and with the person lying in the left lateral position.

In **aortic regurgitation**, the characteristic murmur is a diastolic blowing decrescendo murmur best heard at the left sternal border with the person sitting up and leaning forward in full expiration.

In **aortic stenosis** the characteristic murmur is a loud, low pitched mid-systolic ejection murmur best heard in the aortic area, radiating to the neck.

#### 5.1.4 Electrocardiography (ECG) and Chest X-ray (CXR)

All assessments of a new murmur or established rheumatic heart disease should include ECG and CXR. ECG is essential to determine rhythm. The CXR helps to assess the size of the heart chambers and to detect pulmonary congestion.

### 5.1.5 Echocardiography <sup>(12)</sup>

All persons with murmurs suggestive of valve disease, or a past history of ARF, should have an echocardiogram. Echocardiography will detect any rheumatic valve damage, help determine its severity and assess left ventricular function. Regular echocardiography helps to detect evidence of progression of valve lesions over time and to assess heart function before surgery.

## 5.2 Management of RHD <sup>(4, 12)</sup>

The management of RHD is complex and requires careful co-ordination. The main goal is to prevent disease progression and to avoid, or at least delay, valve surgery. Secondary prophylaxis for prevention of recurrent ARF is the main strategy to achieve this (refer to *4 Secondary Prophylaxis*). Regular clinical review is essential and follow-up echocardiography is important to follow the progress of the heart valve lesions. Management of RHD depends on the severity of disease. Basic guidelines for management of mild, moderate and severe RHD are provided below.

The key elements in the effective management of RHD are:

- Initial assessment, education and referral to a medical or heart specialist
- Management of heart failure (diuretics and ACE inhibitors)
- Management of atrial fibrillation (Digoxin and anti-coagulation)
- Regular medical review and carefully planned follow-up
- Regular secondary prophylaxis (to prevent recurrent ARF)
- Infective endocarditis prophylaxis before dental or surgical procedures
- Regular dental care
- Family planning referral (for women)
- Appropriate surgical discussions and intervention
- Special considerations (e.g. managing RHD during existing pregnancy)
- Influenza and pneumococcal vaccination (where available)

## 5.3 RHD and Pregnancy

Pregnancy causes stress to the heart and may make any existing valve problem worse. The cardiovascular changes which occur during pregnancy in women with RHD may threaten the health of the woman and the foetus. Changes that occur during pregnancy are

- increased heart rate and blood volume
- reduction in systemic and pulmonary resistance
- increased cardiac output.

These changes can complicate existing valvular heart disease and may cause life-threatening complications during pregnancy. Sub-clinical RHD may be identified for the first time during pregnancy because of the above changes. Women with RHD are at high risk of complications immediately after birth.

Ideally, women with known rheumatic valve disease should be fully assessed before pregnancy occurs so that any necessary intervention may safely occur. Women at particular high risk may be counselled to avoid pregnancy (e.g. severe pulmonary hypertension). When pregnancy occurs, management depends on the type and severity of heart valve disease. It is essential that a pregnant woman be assessed by a medical specialist as early as possible so that a coordinated pregnancy management and follow-up can be planned. Management generally includes:

- restricting physical activity and salt intake;
- administering appropriate secondary prophylaxis;
- avoiding community-acquired infectious diseases;
- education about monitoring own signs and symptoms and seeking care if shortness of breath;
- close monitoring of cardiovascular state (specifically in woman who have symptoms of RHD).

Special attention should be given to women with high risk RHD including women with

- mitral and/or aortic stenosis;
- atrial fibrillation;

- prosthetic heart valves;
- those receiving anticoagulant therapy with warfarin

#### 5.4 Endocarditis prevention

Infective Endocarditis is a serious complication of RHD and may also occur after heart valve surgery. Endocarditis is caused by bacteria in the bloodstream. This uncommonly occurs during dental or surgical procedures but often the source of the infection is not clear. In people with rheumatic valve disease, endocarditis most commonly occurs in the mitral or aortic valves since these are the most commonly damaged heart valves. Although the effectiveness of prophylactic antibiotics before dental or surgical procedures has not been proven to reduce the likelihood of developing endocarditis, they have been traditionally given as a preventive measure. Therefore, people with RHD or artificial heart valves should receive antibiotics before procedures that may introduce bacteria into the bloodstream.

People with a history of ARF but no valve damage do not require antibiotic prophylaxis before procedures. People who receive regular Benzathine penicillin or oral Penicillin for secondary prophylaxis should be offered a different antibiotic to prevent endocarditis (see Table 7 and Table 8 below).

**All people with ARF and RHD should have regular dental care to prevent dental decay and the potential risk of endocarditis.**

DENTAL PROCEDURES	OTHER PROCEDURES
Dental extractions	Tonsillectomy/adenoidectomy
Periodontal procedures	Bronchoscopy with a rigid bronchoscope
Dental implant placement	Surgery involving the bronchial mucosa
Gingival surgery	Sclerotherapy of oesophageal varices
Initial placement of orthodontic appliances	Dilatation of oesophageal stricture
Surgical drainage of dental abscess	Surgery of the intestinal mucosa or biliary tract ( <i>except for endoscopy, biopsy and percutaneous endoscopic gastrostomy</i> )
Maxillary or mandibular osteotomies	Endoscopic retrograde cholangiography
Surgical repair or fixation of a fractured jaw	Prostate surgery
Endodontic surgery and instrumentation	Cystoscopy and urethral dilatation
Intra-ligamentary local anaesthetic injections	Vaginal delivery in the presence of infection, prolonged labour or prolonged rupture of membranes
Dental cleaning where bleeding is expected	Surgical procedures of the genitourinary tract in the presence of infection ( <i>e.g. urethral catheterisation, uterine dilatation and curettage, abortion, sterilisation, placement or removal of intrauterine contraceptive devices</i> )
Placement of orthodontic bands	

**Table 6.** Procedures for which Endocarditis prevention is recommended

Situation	Antibiotic	Dose
Standard oral	Amoxicillin	1 dose
Parenteral	Ampicillin	1 dose (IV or IM)
Penicillin allergy	Clindamycin	1 dose
Oral	Cephalexin / Cefadroxil	1 dose
Parenteral	Cefazolin	1 dose

**Table 7.** Suggested prophylactic antibiotic regimens for dental, oral, respiratory tract and oesophageal procedures

Situation	Antibiotic	Dose
High risk	Amoxicillin AND Gentamycin	2 Doses
High risk / penicillin allergy	Vancomycin AND Gentamycin	1 Dose
Moderate risk	Amoxicillin <i>or</i> Ampicillin	1 Dose
Moderate risk / penicillin allergy	Vancomycin alone	1 Dose

**Table 8.** Suggested prophylactic antibiotic regimens for gastrointestinal and genitourinary tract procedures

## 5.5 Surgery for rheumatic heart disease

Surgery may be necessary for severe chronic rheumatic valve disease. However, it is rarely required acutely for carditis associated with acute rheumatic fever. The need for surgery is determined by the severity of symptoms, evidence that the heart valves are significantly damaged and LV chamber size and function. Surgery to repair or replace damaged heart valves is important to prevent left ventricular dysfunction and severe pulmonary hypertension.

### 5.5.1 Assessment for surgery

Echocardiography before surgery helps to assess the severity of valve disease and left ventricular function. If echocardiography is not available, a diagnosis of valve disease must rely on careful clinical examination, ECG and chest X-ray before an individual is referred to a cardiac surgical centre. The results of surgical treatment depend on the following:

- severity of the valve disease at the time of surgery
- left ventricular function
- nutritional status of the individual prior to surgery
- long-term post-operative management (particularly anticoagulation management).

Impaired left ventricular function, atrial fibrillation, diabetes and other co-morbidity can all increase surgical risk and decrease long-term survival rates after surgery. People who require emergency surgery or re-operations have an increased morbidity and mortality following surgery.

### 5.5.2 Contra-indications to surgery

There are very few absolute contra-indications to valve surgery; each person must be assessed individually to determine the risks and benefits of surgery. Factors such as poor left ventricular function with valve regurgitation, severe pulmonary hypertension may pose an unacceptable risk for cardiac surgery.

Age of the person and the presence of co-morbidities also affect the risk/benefit of surgery. Young people often recover well after surgery, even from severe valve disease. Co-morbidity has a much more pronounced effect in older people. Co-existing health problems which require consideration before surgery include:

- chronic kidney disease ;
- chronic obstructive pulmonary disease;
- cerebral and peripheral vascular disease;
- cancer;
- obesity.

Good nutritional status improves post-operative outcomes.

### 5.5.3 Long-term complications

Many of the long-term complications following valve surgery are related more to the individual and the quality and frequency of management rather than to the surgical procedure or prosthesis. Complications may include prosthetic valve thrombosis and degeneration, thromboembolism, endocarditis and bleeding.

### 5.5.4 Long-term postoperative management

Heart valve surgery requires regular long-term follow-up. Ideally, this should be done in a centre equipped with echocardiography. Individuals who have had conservative valve procedures, such as valvotomy or valve repair, require close observation to detect re-stenosis or a recurrence of valve regurgitation, and to ensure secondary prophylaxis is administered regularly. It is also important to monitor LV and prosthetic function.

If echocardiography is not available, patients should be referred back to the surgical centre or local medical specialist if they develop any of the following:

- recurrent symptoms
- evidence of heart failure
- a new regurgitant murmur
- any thromboembolic episode
- signs and symptoms suggesting endocarditis.

Anticoagulation needs to be monitored following replacement with mechanical valves. Good anticoagulation management requires standardized anticoagulation measurement, using the International Normalised Ratio (INR);

The dose of anticoagulation required depends on the individual and the type of prosthetic heart valve used. Regular monitoring of the INR and maintaining it within the therapeutic range may be difficult for people living in areas where health services are difficult to access.

**All people requiring anticoagulation must receive regular care from a Medical Specialist**

Classification	Criteria	Review and Management Plan	Frequency
Low Risk (Priority level 3)	ARF with no evidence of RHD <i>or</i> Trivial to mild valvular disease	Secondary prophylaxis	4-weekly
		Doctor review	Yearly
		Dental review	Yearly
		Echocardiogram (if available)	Children 2-yearly Adults 2-3 yearly
Medium Risk (Priority level 2)	Any moderate valve lesion in the absence of symptoms and with normal left ventricular function <i>or</i> Mechanical prosthetic valves	Secondary prophylaxis	4-weekly
		Doctor review	6-monthly
		Influenza vaccination	Yearly
		ECG (optional)	Yearly
		Medical or Heart specialist review	Yearly
		Echocardiogram	Yearly
		Dental review	Yearly
		Polysaccharide pneumococcal vaccination	5-yearly (max 3 doses)
		Endocarditis prevention	As required
High Risk (Priority level 1)	Severe valvular disease, or Moderate/ severe valvular disease with symptoms, or Tissue prosthetic valves and valve repairs	Secondary prophylaxis	3-4 weekly
		Doctor review	3-6 monthly
		Influenza vaccination	Yearly
		ECG (optional)	Yearly
		Medical or Heart specialist review	3-6 monthly
		Echocardiogram	3-6 monthly
		Dental review	Within 3 months and yearly thereafter
		Polysaccharide pneumococcal vaccination	5-yearly (max 3 doses)
		Endocarditis prevention	As required
		Warfarin + Aspirin	As prescribed

**Table 9.** Recommended routine review and management plan for ARF and RHD <sup>6</sup>

## 6 GUIDELINES FOR NOTIFICATION AND DATA MANAGEMENT

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### 6.1 Introduction to Disease Registers

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

**Safe** so that it is not damaged or lost, and

**Secure** to maintain privacy

#### 6.1.1 Paper Register (Book or List)

Simple paper registers may contain information for a local health facility, or may provide information to a larger, central computer register. Examples of paper registers include

- Benzathine Penicillin G injection book (names, contact information, dates injections given and next due)
- Cardiac surgery list (names, and dates and outcomes of surgery)

Potential problems with paper registers:

- The information may not be secure - pages may be torn from a book or damaged by water;
- The same person may be entered more than once - the person could accidentally be counted more than once resulting in incorrect reporting;
- Long handwritten lists may be difficult to search and analyse - this may result in incorrect reporting;
- Important information may be missing - this may result in incorrect health management and/or incorrect reporting.

Benefits of paper registers:

- They can be easily transported;
- Usually no special training is required for their use;
- Useful where computers are not available or where computer support is difficult.

#### 6.1.2 Computer Register (Database)

A computer registers is called a *database*. A database is able to record specific information for many people. Potential problems with computer registers:

- Expense of purchase and maintenance, and security of equipment
- Need for training and support
- Electricity interruptions

Benefits of computer registers:

- The information is safe - passwords and computer back-up protect the information;
- Each person is only entered once - the computer can be set up to prevent duplicate record entry;
- Information can be searched and sorted quickly;
- Large amounts of information can be changed or updated quickly if required;
- Reports can be produced accurately and automatically.
- They can be easily backed up and stored in a number of places so that data is not lost if the main computer fails.

#### 6.1.3 Information on the RHD Register

The following information should be included on an RHD Register. All information could be stored on a centralised register, or some information (e.g. individual doses of Benzathine penicillin G) could be entered onto local Benzathine lists only.

- Unique identification. This may be a clinic or hospital number.
- Personal information including name/s, date of birth, gender, ethnicity, contact information
- ARF diagnoses, date of diagnoses, where the diagnosis was made, presence of chorea
- Current disease status / RHD priority (refer to 6.2 *Management of RHD*)
- Secondary prophylaxis details (medication, dose, frequency, date started, expected cease date and actual cease date, and number of Benzathine penicillin injections received)

- Surgery details (surgical procedures and surgery dates)
- Dates for next review with medical specialist or echocardiogram (or date for heart valve surgery)
- Date and cause of death.

Other information on the register might include

- Benzathine penicillin injections – dates given and date due
- Echocardiogram – dates done and results
- Specialist medical review and dental review – dates of reviews, clinical findings, management.

**An RHD register should be simple to use AND contain enough information to provide adequate clinical management and reporting.**

## 6.2 Notification

ALL cases of confirmed OR suspected ARF and RHD should be notified to the RHD register (or local health authority, if applicable) to

- Identify high risk individuals who require priority care
- Assist health staff to coordinate secondary prophylaxis and follow-up care
- Help to identify others in the family or community who may be at risk
- Provide information on the local rates of disease.

Notification should include

- Name, date of birth and contact information
- Hospital Number or clinic number
- Diagnosis (e.g. ARF, RHD, unconfirmed ARF) and date of diagnosis

**CHECK LOCAL GUIDELINES:  
Consent may be required to record personal information on a disease register.**

## 6.3 Data Management

### 6.3.1 Sources of Information

Finding and recording cases of ARF and RHD may be demanding in the early stage of establishing a register. The following may help to find large numbers of cases quickly:

- Benzathine Penicillin G injection lists/ books maintained in clinics and treatment rooms;
- Cardiac surgery lists;
- Echocardiogram lists, books and reports;
- Patient lists from cardiologists, physicians, dentists and researchers;
- Hospital admission / discharge reports including ICD-9 or ICD-10 coding
- School screening referrals

### 6.3.2 Reports from the Register

Based on the information listed in 6.1.3 above, a number of reports can be produced from the register. These include

- Basic epidemiology (refer to *9 Programme Evaluation and Reporting*)
- Lists of individuals needing urgent care – high priority RHD cases;
- Lists of people receiving inadequate secondary prophylaxis;
- Delivery of Benzathine penicillin injections;
- People on Specialist review lists;
- People on Echocardiogram and Surgery waiting lists;
- People who are deceased and their cause of death.

## 7 RHD CONTROL PROGRAMMES

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### 7.1 Elements of the Programme <sup>(4)</sup>

The recommended elements of a national RHD programme are based on the World Health Organisation principles and include:

- A strong commitment from local Government (Ministries of Health and Education);
- A skilled and committed RHD advisory committee established within the Ministry of Health, with membership from medical and nursing professionals, educators, and community groups
- An RHD Register of all individuals with confirmed and suspected ARF and RHD
- Secondary prevention activities integrated into existing health care systems and aimed at preventing the recurrence of ARF and severe RHD;
- National notification of ARF and RHD to the Ministry of Health.
- A priority system to help deliver services to those at greatest risk
- Gradual programme establishment, starting in a central area and extending to regional and national coverage
- Reliable resources including medication supply and support from a microbiology laboratory

### 7.2 Core Programme Objectives

An RHD control programme should aim to improve early diagnosis of, and standardise care for people with ARF and RHD. Core programme objectives should be to:

- Identify and register cases of known (or suspected) ARF and RHD;
- Standardise, monitor and improve delivery of secondary prophylaxis;
- Standardise diagnosis and management of ARF and RHD;
- Provide training and support for health workers (especially those in rural and remote areas);
- Support education at community level;
- Report on the programme and rates of disease.

#### 7.2.1 Registering cases of ARF and RHD

In the early stages of identifying and registering people with ARF and RHD, the following sources may contain information on known or suspected cases:

- Benzathine penicillin injection lists
- Cardiac surgery lists;
- Echocardiogram lists and reports;
- Lists from cardiologists, physicians and researchers;
- Hospital admission reports;
  - Admission books
  - ICD-9 or ICD-10 coding from hospital computer systems.

People who present to hospitals and health facility over time should be reported (notified) to the RHD programme (and to the local health authority, if applicable).

### 7.3 Secondary Prophylaxis <sup>(4, 12)</sup>

#### 7.3.1 Monitoring secondary prophylaxis

Recurrent ARF is almost always a result of failing to receive adequate secondary prophylaxis. It is thought that poor delivery of secondary prophylaxis is caused by a number of factors, some of which are listed below.

**1. The relationship** between the person requiring prophylaxis medication and staff at the health facility. It has been reported by some communities that people are more likely to present to a health facility where there is familiar and competent staff who provide a caring and holistic health service.

**2. The level of education and training.** Health workers and/or people with ARF/ RHD may not fully understand the role of secondary prophylaxis in preventing ARF and subsequent heart damage.

3. **Refusal.** Some people do not want to receive treatment despite their level of understanding.
4. **Difficulties accessing health care.** Traveling to the health facility to receive treatment may be difficult and/or costly, especially for people living in rural and remote areas.
5. **Forgetting to attend** the health centre on the date when secondary prophylaxis is due.
6. **The pain** associated with Benzathine penicillin injections is a barrier to receiving treatment for some people.
7. **Staff workloads and priorities.** Health staff may be unable to identify and encourage people who do not receive regular secondary prophylaxis. Other serious health issues may compete for time and resources.
8. **Cost of treatment** (where individuals are required to pay for health care and penicillin)

Communication with regional health facilities will help identify people who are not receiving adequate secondary prophylaxis. Together, an RHD control programme and treating health staff can

- Develop ways to improve uptake of secondary prophylaxis at a local level
- Provide education for people who do not receive treatment
- Notify clinicians of those who do not receive secondary prophylaxis and are at increased risk of ARF recurrence so that they can be monitored closely.

### 7.3.2 Measuring Benzathine Penicillin injection delivery

People who do not receive adequate secondary prophylaxis should be identified and monitored. Monitoring Benzathine penicillin injection delivery is important to ensure that individuals receive adequate treatment to help prevent recurrent ARF.

Secondary prophylaxis delivery can be monitored by

- Identifying people who are late for a scheduled BPG injection or who regularly fail to receive Benzathine penicillin G injections
- Identifying people or who occasionally miss twice daily tablets or who regularly fail to renew supplies of oral medication.

Percent of Benzathine penicillin injections delivered is calculated from the total number of injections required:

- People prescribed **3-weekly** injections should receive **17** injections in 12 months
- People prescribed **4-weekly** injections should receive **13** injections in 12 months.

To calculate the percent of injections received for an individual:

- *Record* the number of injections PRESCRIBED for a full 12 months (e.g. the number of injections prescribed from January and December 2007 for a person on 4-weekly treatment = **13**)
- *Count* the number of injections RECEIVED during the 12 months (e.g. **10** injections may have been received)

*Calculate:* number of injections RECEIVED (**10**) divided by the number PRESCRIBED (**13**) and multiply by **100**  
 $(10 \div 13) \times 100 = 77\%$

In this example, the person received **77%** of prescribed injections in 2007.

#### **NOTES:**

***Receiving less than 80% of injections places an individual at a higher risk of recurrent ARF. Follow-up may be required.***

***If injections were prescribed for the full year but no injections were received, record 0%.***

To measure the percent of injections received by a group of people at a specific health facility:

- *Record* the total number of injections PRESCRIBED for a full 12 months (e.g. if 12 people require 4-weekly injections, this would be 12 (people) multiplied by 13 (injections) = **156 injections**.)
- *Count* the total number of injections RECEIVED by all people in the full year (e.g. a total of **124 injections** may have been given to the 12 people)

*Calculate:* total number RECEIVED (**124**) divided by the total number PRESCRIBED (**156**) and multiply by **100**  
 $(124 \div 156) \times 100 = 86\%$

Overall Benzathine penicillin injection delivery for this health facility in 2007 is **86%**.

***NOTE: Include only those persons prescribed Benzathine penicillin injections for the FULL 12 months.***

### 7.3.3 Strategies for improvement

A dedicated health worker(s) at each health facility should be nominated to work closely with other staff and people requiring secondary prophylaxis. Strategies to improve service delivery include

#### 1. Identifying people who require secondary prophylaxis.

- Each health centre should maintain a list of people who attend the health service and require secondary prophylaxis to prevent ARF.

#### 2. Disease education for people requiring treatment and their families

- Delivery of secondary prophylaxis presents regular opportunities to provide education and encouragement to individuals and their families to continue to attend for treatment.

#### 3. Establishing systems to recall people who miss doses.

- Clinic cards, reminder notes, and communication with parents and teachers may help reduce the number of missed doses.
- Negotiating times such as 'on the way to school (or work) on Mondays' may be useful for people who forget their secondary prophylaxis.
- A weekly clinic dedicated to people requiring secondary prophylaxis will help standardise appointment times and reduce the day-to-day burden on health staff. (*people who present for treatment outside set clinic times should not be turned away*)

#### 4. Focusing on individuals who continue to default treatment.

- The secondary prophylaxis list should be consulted regularly to identify people who are late for injections.
- Improve the quality, frequency and delivery of health education.
- Identify and educate others who can provide support and encouragement for people who regularly miss treatment.

#### 5. Communicating about people who move between communities.

- Health facilities should communicate where possible about people who move between locations and require regular medication (e.g. children who attend boarding school, people visiting other communities, prison inmates, extended holiday or work transfer)

#### 6. Promote strategies to reduce injection pain.

- See 5.7.4 Pain Reduction

### 7.3.4 Antibiotic Supply

A reliable local supply of secondary prophylaxis antibiotics (particularly Benzathine penicillin G) is essential to RHD control. If Benzathine penicillin G supplies are low or not available, the following steps should be taken:

- Confirm when Benzathine penicillin G 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 Benzathine penicillin G is readily available.

Health staff should clearly inform people who usually receive Benzathine penicillin G that oral penicillin is only a temporary solution, and that they will be recalled when the Benzathine penicillin G injections become available.

## 7.4 Training and Support for Health Workers

### 7.4.1 Standard Care

Guidelines for diagnosis and management of people with ARF and RHD should be based on locally approved guidelines, which may be adapted from the WHO Technical document <sup>(4)</sup> and/or other sources.

### 7.4.2 Training

RHD workshops for qualified health staff help to develop skills required for diagnosis and management of ARF and RHD at the primary health care level. The RHD Programme should also contribute to existing health worker training programmes including:

- Student Nurse training

- Ongoing education for trained nurses
- Nurse practitioner curriculums
- Medical student training
- Medical Attendant training
- Orientation of new staff

Delay in diagnosis of ARF or missed diagnosis can result in high rates of RHD in the community and the need for complex tertiary care. Specifically, training for health staff should focus on

- Identifying and monitoring children at high risk of developing ARF;
- Observing for signs and symptoms of ARF and referring for medical assessment;
- Delivering secondary prophylaxis effectively to prevent recurrent episodes of ARF;
- Providing disease education to individuals and families in their communities;
- Communicating regularly with the RHD Programme about people with ARF and RHD under their care.

**A questionnaire can help determine participant's level of knowledge, and help evaluate the effectiveness of workshops.**

### **7.4.3 Communication**

The RHD Programme should provide ongoing support for health workers, especially those in rural and remote areas. Support may include

- Discussion about new and existing cases of ARF and RHD in the area;
- Updated policies to enhance clinical care ;
- Regular reports from the programme about;
  - The impact of ARF and RHD in the community,
  - The effectiveness of control strategies in reducing the burden of heart disease.

## **7.5 Community Support**

### **7.5.1 Education**

People with ARF and RHD and their families should receive detailed education – ideally through their local health staff - about the progression of disease, the importance of secondary prophylaxis, and the risk and outcomes of moderate or severe heart disease. Education should be conducted at diagnosis, during hospital admissions and when presenting for secondary prophylaxis and in preparation for cardiac surgery. Education aims to

- Provide information about ARF and RHD and the personal and relevant risks to the individual associated with worsening heart disease;
- Help individuals recognise their own signs and symptoms of ARF and present to health services for assessment;
- Focus on the importance of secondary prevention;
- Include instructions for further information and ongoing care.

### **7.5.2 Referral**

Following identification of ARF or RHD it is important that communication is established between the hospital and relevant local health facility to:

- Discuss ongoing clinical management
- Optimise delivery of secondary prophylaxis
- Recall for specialist review or surgery

A referral letter following hospital discharge provides a written record that can be stored in the local health facility medical file for future reference. Communication may be difficult in some areas, especially in the absence of reliable telephone services. Existing communication networks with rural and remote areas should be used where possible (e.g. with regular pharmacy/ hospital supplies, remote specialist or allied health visits, during health staff training programmes).

## **7.6 Screening for RHD**

Priority should be given to identifying acute cases early in the programme. Screening may be undertaken later if time and resources become available. Screening for RHD is valuable in areas where a large number of cases are expected to be found, however the following should be considered:

- Who will be screened (e.g. school children are easiest to screen, although RHD rates may be higher in young adults)
- Methods of diagnosis available (e.g. auscultation & clinical assessment, echocardiogram)
- The availability of trained staff (e.g. echocardiogram technicians, clinicians)
- A process for reporting results to the RHD register (e.g. data collection forms)
- Health resources available for long-term management of more RHD cases.

## **7.7 Programme Difficulties**

Maintaining a dedicated RHD programme may be difficult, especially if the programme does not work well within the local health system. The most common reasons for programme failure include

- A lack of local resources to maintain the programme
- Limited funding agreements
- Heavy workloads for primary care health staff
- Other health issues demand available resources (Tuberculosis, Malaria, HIV/AIDS, respiratory disease)
- The RHD programme setup is too complex
  - Communication with rural and remote health facilities is difficult
  - Travel to provide health worker training may not be available
  - Data management may be too demanding
  - The programme may be expanded to quickly

## **7.8 Programme Sustainability**

The following points should be considered for a sustainable RHD programme:

- Dedicated, long-term commitment and funding from the Ministry of Health
- A dedicated and skilled RHD programme coordinator
- Integration with existing programmes (e.g. Tuberculosis DOTS nurses also responsible for coordinating secondary prophylaxis)
- Existing systems used to identify and manage of ARF and RHD
  - School health / screening programmes
  - Disease notification forms and notification process
  - Medical specialist referral process
  - Drug supplies to remote areas
  - Training programmes for health staff

## 8 PROGRAMME EVALUATION AND REPORTING

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### 8.1 Programme Evaluation

A successful RHD programme can demonstrate lowering rates of recurrent ARF; however the number of people with RHD may continue to rise as existing cases in the community are found.

Evaluating the programme and reporting results will:

- Help describe ARF and RHD in the community
- Demonstrate whether control strategies are effectively reducing the burden of RHD
- Assist local health authorities with future programme planning and resource allocation

Programme evaluation should include disease epidemiology and standards of clinical care.

#### 8.1.1 Epidemiology

The following information should be reported for a specific time frame (e.g. the previous 12 months):

- The number of people diagnosed and registered with ARF and RHD during the previous year;
- Percent (%) of total reported ARF episodes that were recurrences;
- Age- specific and overall incidence of ARF;
- Age-specific and overall point prevalence of RHD;
- Percent (%) of newly registered individuals diagnosed with RHD (i.e. with a history of ARF that has been missed);
- Percent (%) of RHD cases which are classified as moderate to severe;
- The number of deaths directly resulting from RHD (by age group).

#### 8.1.2 Standards of clinical care

- Secondary prophylaxis delivery
  - The total number of BPG injections given as a proportion (%) of those scheduled to be given over 12 months
  - Percent (%) of individuals who received 80% or more of their Benzathine penicillin injections
  - Percent (%) of individuals who received 50% or less of their Benzathine penicillin injections
  - Percent (%) of individuals who do not receive any prescribed Benzathine penicillin injections
- The level of secondary prophylaxis delivery by health facility
  - Percent (%) of health facilities that delivered
    - 80% or more of the scheduled Benzathine penicillin injections
    - 50% or less of the scheduled Benzathine penicillin injections
- The number of heart operations (in children)

### 8.2 Programme Report

The RHD Programme report should include evaluation results (above), programme activities and outcomes, and a financial budget.

#### 8.2.1 Suggestions for reporting

- Epidemiology helps to describe the burden of disease in the community, and identifies age groups which may be most at risk of illness and death.
- Standards of clinical care may also include recent policy changes
- Reporting the number of health workers who have received training will help describe the level of local clinical expertise
- Community education may describe
  - Which materials are used (e.g. posters, pamphlets)
  - How materials have been distributed
  - How education is delivered (e.g. workshops in schools, individual/family sessions)
- How the register is maintained could include
  - How information is reported to the register

- How it is reported back to health care providers (e.g. lists sent to health facilities, medical specialists)
- Any ongoing issues with data management
- Roles and responsibilities of the RHD advisory committee
- Funding: sources and how it is used, including:
  - Staff wages
  - Office and communication supplies
  - Travel and accommodation costs
  - Education materials
  - Health worker training programmes

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