RHEUMATIC FEVER/ RHEUMATIC HEART DISEASE

Guidelines for Prevention, Diagnosis and Management

Ministry of Health
Jamaica

September 2002

Director, Family Health Services

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FOREWORD

These guidelines for prevention, diagnosis and management of rheumatic fever and rheumatic heart disease, were developed to ensure clarity and consistency in the management of patients with these conditions.

Much input was received from health workers in the field to ensure that all areas of concern were addressed.

These guidelines are expected to be used by medical officers, nurses and nurse practitioners in all types of health facilities.

Quality of care assessments/audits will be done by the Ministry of Health using these guidelines as the basis.

Periodic modifications will be done as necessary.

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I. INTRODUCTION

Rheumatic fever/rheumatic heart disease is still a major public health problem in developing countries. It is the most common acquired cardiovascular disease in children and young adults, affecting some 12 million people worldwide with about 400,000 deaths annually. The 5– 15 years age group is most often affected but the disease may occur in younger or older individuals as well.

The group A beta-hemolytic streptococcus is the bacterium implicated in the disease. In developing countries where the socio-economic conditions are not satisfactory, the occurrence of Rheumatic Fever/Rheumatic Heart Disease is high with mortality rates up to 8 per 100,000 population, prevalence rate up to 10 per 1,000 school age children and incidence rate up to 100 per 1,000 school age children. In developed countries, its occurrence is nil or very low. In developing countries, the disease has a high rate of recurrence and severity. It also accounts for 12– 50% of all cardiac patients admitted to hospitals. Rheumatic heart disease is a chronic disease, which requires repeated hospitalizations and surgery and often leads to premature death and years of productive life lost.

Rheumatic fever and rheumatic heart disease require enormous resources for the medical and surgical treatment of patients, many of whom may have reduced productivity and eventually die. A hospital survey done in Jamaica in 1995 revealed a cost of JA$105,000 per admission of 21 days per case and an estimated annual expenditure of JA$17million in just three hospitals (KPH, University and Bustamante hospitals)

Almost half of rheumatic fever/rheumatic heart disease patients are unaware of their disease and only about two thirds of patients receive monthly secondary prophylaxis. In the absence of an anti-streptococcal vaccine, there are feasible and proven cost-effective methods of prevention including the adequate management of sore throats.

This document serves to guide health care workers in the adequate prevention, diagnosis and management of rheumatic fever and rheumatic heart disease including the programme requirements for reporting and investigating. The document should be used as such and shared with all health workers involved in the management of such cases.

II. PRIMARY PREVENTION OF RHEUMATIC FEVER

II.1. MANAGEMENT OF SORE THROATS

INTRODUCTION

Group A beta-haemolytic streptococcal infections of the upper respiratory tract in particular the throat, are responsible for initial and recurrent attacks of rheumatic fever. Group A streptococcal infections are universally endemic and currently there is no vaccine to prevent these infections.

Prevention of rheumatic fever therefore depends on the accurate clinical diagnosis and management of sore throats using the most appropriate antibiotic treatment.

EPIDEMIOLOGY

Humans are the only significant reservoir for the Group A streptococcus organism. Children between the ages of 5 to 15 are most susceptible but all ages may be affected. Residential institutional settings such as schools and military barracks are usually the sites of epidemics where crowding facilitates the spread of infection by droplets.

There are about 100 recognized serotypes of group A streptococci. Infection with one confers long lasting type specific immunity but because of the abundance of stereotypes, the risk of new infection is continuous. The more "rheumatogenic" stereotypes are most often associated with upper respiratory tract infections hence rheumatic fever usually follows upper respiratory tract infections and not streptococcal skin infections.

MANAGEMENT OF GROUP A STREPTOCOCCAL UPPER RESPIRATORY TRACT INFECTION

An accurate clinical diagnosis of group A streptococcal upper respiratory tract infection may be difficult to make and may require the support of laboratory investigations (throat swab). However if unable to do cultures, use the clinical features and treat accordingly. If in doubt, treat as a 'strep' throat. The typical sore throat presents with sudden onset of high fever (greater than 38°C), pain on swallowing, tenderness in anterior cervical area and maybe accompanied by abdominal pain, nausea and vomiting. Younger children (<3 years) usually present with lower grade fever, irritability and a serous discharge from the nostrils. Anterior cervical lymphadenitis is also a characteristic finding. On examination, the throat appears red with exudates on the pharynx or tonsils. Punctate haemorrhages may be on the soft palate. See Table 1 for comparison of clinical features with "non-strep throat".

A throat swab of the posterior pharynx, tonsils or tonsillar fossae should be done and sent for culture to confirm group A streptococcus. If unable to do the throat swab, it is best to treat as a group A streptococcal infection on clinical features. Group A streptococcal antibody tests such as the antistreptolysin O (ASO) and the anti-deoxyribonuclease B (anti-DNase B) are not useful for diagnosing acute group A streptococcal pharyngitis as they may be elevated in normal healthy school children and the antibody response occurs many days after the onset of infection.

Table1: Comparison of Clinical Characteristics of "Strep" and "Non-Strep" Throats

Clinical Characteristics	Strep Throat	Non-Strep Throat
AGE	5-15 Years (most common)	All ages
MODE OF ONSET	Sudden	More gradually
INITIAL SYMPTOMS	Sore throat with pain while swallowing	Mild sore throat
FEVER	High (over 38°C)	Not so high
APPEARANCE OF THE THROAT	• Redness, hyperemia, oedema and exudates (yellow flecks) of the pharynx* • Enlargement of the tonsils with exudate* • Hyperemia, oedema and punctate haemorrhages in the soft palate*	Redness of the pharynx
OTHER SIGNS	• Tenderness of the anterior cervical lymph nodes* • Scabby erosions on the edges of the nostrils* • Clinical picture of Scarlet Fever°*	• Cough + • Hoarseness + • Watery nasal secretion + • Conjunctivitis +

* Characteristic manifestation of a "strep" throat

\+ Non-characteristic manifestation of a "strep" throat

° Red strawberry tongue, cutaneous eruption particularly on the throat, chest, axillae, elbows, groins and inner surface of the thighs.

The recent onset of the characteristic clinical picture with some of the frequent characteristic manifestations of a "strep" throat (*) and none of the non-strep throat manifestations (+) makes a clinical diagnosis of streptococcal pharyngitis or tonsillitis likely.

ANTIMICROBIAL TREATMENT FOR GROUP A STREPTOCOCCAL INFECTION

Penicillin remains the treatment of choice for group A streptococcal upper respiratory tract infections and in fact has been demonstrated to prevent rheumatic fever. Cefalosporins are also capable of eradicating the organism from the upper respiratory tract but are more expensive than penicillin.

A single intra-muscular injection of benzathine benzylpenicillin is the most effective treatment in eradicating group A streptococci due to its long duration of action. For persons less than 30 kg in weight, the dose is 600,000 IU. For 30 kg and over the dose is 1,200,000 IU.

If oral penicillin is to be used, the duration of treatment should be no less than 10 days. For persons allergic to penicillin, oral erythromycin for 10 days is the treatment of choice. See Table 2. Newer macrolides may also be used but are more expensive.

After a full course of antibiotic therapy there is no need to re-culture patients. If still symptomatic after a complete course of antibiotics in which compliance has been assured, then give a second course of antibiotics in adequate dosage. A repeat of the first course may be used, i.e. the same drug. (These instances are rare and laboratory documentation of the organism may be advantageous to monitor treatment.)

Table 2: Treatment of Group A Streptococcal Pharyngitis
(Primary Prevention of Rheumatic Fever)

No bactrim

ANTIBIOTIC	ROUTE	DOSE	DURATION OF RX
For non-penicillin allergic patients: **Benzathine benzylpenicillin**	**IM**	<30 kg 600,000 IU ≥30 kg 1,200,000 IU	A single injection
Phenoxymethylpenicillin Pen V Amoxicillin 50mg/kg/day	**Oral**	<30 kg 250 mg 2 or 3 times daily ≥30kg 500 mg 2 or 3 times daily	10 days
For penicillin allergic patients:			
Erythromycin ethylsuccinate	**Oral**	40 mg/kg/day (max. 1.5 g/day) 3 times daily	10 days
Erythromycin estolate	**Oral**	20– 40 mg/kg/day (max. 1.5 g/day) 3 times daily	10 days

Cephalosporins; Azithromycin 12mg/kg/Day for 5 days

Guideline: MANAGEMENT OF SORE THROATS		
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III. DIAGNOSIS AND MANAGEMENT OF RHEUMATIC FEVER

1. Diagnosis of Rheumatic Fever
2. Management of Rheumatic Fever

III.1. DIAGNOSIS OF RHEUMATIC FEVER

Hospitalization

Rheumatic Fever is an inflammatory, non-suppurative sequel of Group A Streptococcal upper respiratory tract infections with a marked tendency to recur. It affects mainly the large Joints (**Arthritis**) and the Heart (**Carditis**) and less frequently the Brain (**Chorea**), Skin (**Erythema Marginatum**) and subcutaneous tissues (**Subcutaneous Nodules**).

The child with Rheumatic Fever usually has fever and joint pains and swelling, involving several joints (migratory polyarthritis). The larger joints (knees, ankles, wrists or elbows) are more commonly affected and there is frequently more pain than swelling. Migratory polyarthritis is the rule (i.e. more than one joint is affected and their involvement tends to overlap in time such as one knee may become painful just as the other knee or ankle previously inflamed is beginning to improve).

Chorea is purposeless, involuntary, rapid movements often associated with muscle weakness and/or behavioural abnormalities. This is usually a delayed manifestation of Rheumatic Fever.

Erythema Marginatum is a distinctive evanescent, pink rash with a pale centre and round or serpiginous margins. The size may vary and they occur mainly on the trunk or proximal extremities, never on the face. The erythema is transient, migratory and may be induced by the application of heat. The rash is non-pruritc, non-indurated and blanches on pressure. Erythema Marginatum is a rare manifestation of Rheumatic Fever.

Subcutaneous nodules are firm, painless nodules, which may present over the extensor surfaces of certain joints such as the elbows, knees and wrists. They may also appear in the occipital region or over the spiny processes of the thoracic and lumbar vertebrae. The overlying skin is mobile and not inflamed. These are rare and most often associated with the presence of Carditis.

Carditis is inflammation of the heart tissues and is virtually always associated with Valvulitis and a significant murmur.

Patients can suffer from dyspnea, tiredness and lack of appetite. Simultaneously, a heart murmur or a pericardial friction rub can appear as a new physical finding or modifying already existing signs of rheumatic heart disease. The most common murmurs of rheumatic fever are those of mitral incompetence, mitral stenosis and aortic incompetence. Choreiform movements are usually obvious when they occur, but can be for a prolonged time misinterpreted as a simple behaviour disturbance.

Rheumatic Heart Disease is the collective term used to refer to the functional and structural alterations of the heart valves affected by rheumatic fever. The heart lesion may cause death or permanent sequelae. The severity of rheumatic heart disease and the prognosis depend on the extent of the carditis and the frequency of recurrent attacks.

Suspicion of Rheumatic Fever is strengthened if the patient has had a sore throat about 2–4 weeks earlier or has laboratory evidence of inflammation or a history of Rheumatic Fever.

These clinical presentations are known as the ***JONES CRITERIA*** and have been recently revised by the World Health Organization (WHO) in 2004 to facilitate diagnostic categories and criteria as shown in the Table below.

Evidence of preceding Group A Streptococcal Infection forms part of the diagnostic criteria for categories 1 and 2 along with two major or one major and two minor manifestations. For category 3 or recurrent attacks of Rheumatic Fever (RF) with established Rheumatic Heart Disease, no further major manifestation is required. However two minor manifestations and evidence of recent/preceding Group A Streptococcal Infection forms part of the diagnosis for Rheumatic Chorea and Chronic Value Lesions, no major or minor manifestations and no evidence of recent Group A Streptococcal Infections are required.

Table 3: 2002-2003 WHO Criteria for the diagnosis of Rheumatic Fever and Rheumatic Heart Disease (based on the revised Jones Criteria[3,4])

Diagnostic Categories	Criteria
1. Primary episode of RF.[a]	Two major* or one major and two minor** manifestations **plus** evidence of a preceding Group A Streptococcal Infection***.
2. Recurrent attack of RF in a patient **without** established Rheumatic Heart Disease.[b]	Two major or one major and two minor manifestations **plus** evidence of a preceding Group A Streptococcal Infection.
3. Recurrent attack of RF in a patient **with** established Rheumatic Heart Disease.	Two minor manifestations **plus** evidence of a preceding Group A Streptococcal Infection[c].
4. Rheumatic Chorea Insidious onset of Rheumatic Carditis.[b]	Other major manifestations or evidence of Group A Streptococcal Infection not required.
5. Chronic valve lesions of RHD (patients presenting for the first time with pure mitral stenosis or mixed mitral valve disease and/or aortic valve disease).[d]	Do not require any other criteria to be diagnosed as having Rheumatic Heart Disease.

*Major Manifestations	- Carditis - Polyarthritis - Chorea - Subcutaneous Nodules - Erythema Marginatum
** Minor Manifestations	- Clinical: Fever, Polyarthralgia - Laboratory: Elevated acute phase reactants (Erythrocyte sedimentation rate or Leukocyte count)
*** Supporting evidence of a preceding streptococcal infection within the last 45 days	- Electrocardiogram: prolonged P-R interval - Elevated or rising Antistreptolysin-O or other streptococcal antibody, or - A positive throat culture, or - Rapid antigen test for Group A Streptococci, or - Recent Scarlet Fever

[a] Patients may present with polyarthritis (or with only polyarthralgia or monoarthritis) and with several (3 or more) other minor manifestations, together with evidence of recent Group A Streptococcal Infection. Some of these cases may later turn out to be Rheumatic Fever. It is prudent to consider them as cases of "Probable Rheumatic Fever" (once other diagnoses are excluded) and advise regular secondary prophylaxis. Such patients require close follow up and regular examination of the heart. This cautious approach is particularly suitable for patients in vulnerable age groups in high incidence settings.

[b] Infective Endocarditis should be excluded.

[c] Some patients with recurrent attacks may not fulfill these criteria

[d] Congenital Heart Disease should be excluded.

DIFFERENTIAL DIAGNOSES OF RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE

Carditis

- Innocent murmurs
- Infective Endocarditis
- Mitral Valve Prolapse
- Congenital Heart Disease
- Viral Carditis

Arthritis

- Infectious (Gonorrhoea)
- Post-infectious (TB, Typhoid)
- Collagen Vascular (Juvenile Rheumatoid Arthritis, SLE)
- Lyme Disease
- Serum Sickness
- Viral Infections (Hepatitis, Rubella, Mumps, Infectious Mononucleosis)
- Haematological (Leukemia, Sickle Cell Disease)

Juvenile Rheumatoid Arthritis (JRA) may be confused if onset is acute polyarthralgia. However in JRA the disease is symmetrical, small joints are involved, the arthritis is less migratory and there is a less dramatic response to ASA therapy. There is also morning joint stiffness, a salmon pink rash and splenomegaly in JRA.

III.2. MANAGEMENT OF RHEUMATIC FEVER

All cases of suspected Rheumatic fever must be referred to hospital for diagnosis and management. There are three main components to the treatment of the acute attack. These are:

(i) Treatment to eliminate the streptococcal infection.

(ii) Treatment for inflammation and for congestive cardiac failure, if present.

(iii) Appropriate rest and supervision

Anti-inflammatory drugs such as aspirin, NSAIDs or steroids should not be commenced until the diagnosis has been established according to the accepted criteria outlined in "DIAGNOSIS". In the interim, acetaminophen may be used for joint pains. Blood cultures should be done to look for evidence of bacterial endocarditis in patients with valvulitis (mitral incompetence and aortic incompetence). The following management guide should be used for all patients:

SPECIFIC MANAGEMENT

1. **Eradicate streptococci as in the treatment outlined in Table 2**

2. **Arthritis**

   - Treat with acetyl salicylic acid (ASA) 80 mg/kg/day X 2 weeks then taper and discontinue over the next 2 weeks

3. **Carditis**

   - BED REST AND DAILY EVALUATION OF THE CARDIOVASCULAR SYSTEM

   Mild Carditis

   - ASA 80– 100 mg X 2–4 weeks then taper and discontinue in 4– 6 weeks

   Moderate to severe or progressive Carditis

   - Prednisone 2 mg/kg/day (max 60 mg/day) for 2– 3 weeks then taper while commencing ASA which is continued for 2– 4 weeks
   - Treatment of cardiac failure with appropriate drugs. Be careful of digoxin in patients with myocarditis.
   - Use captopril in cases with predominant aortic incompetence and/or predominant mitral incompetence

- Add aldactone in severe congestive cardiac failure (monitor serum potassium)

4. Chorea

- No specific treatment. In severe cases, use Haloperidol and nurse in a padded bed.

5. Secondary Prophylaxis

- Commence with intra-muscular benzathine penicillin given every 2 weeks while in hospital. On discharge, give every **28 days.**

6. Notification

- Complete class 1 notification form and investigation form and submit to parish health department or the Surveillance Unit **within 24 hours of diagnosis on suspicion.**

IV. REPORTING AND INVESTIGATION

1. Reporting
2. Investigation

IV.1. REPORTING

Rheumatic Fever is a class 1 notifiable disease and as such should be reported to the parish health department or the Surveillance Unit within 24 hours of suspicion using a class 1 notification form (see Appendix 1). All areas on the notification form should be completed as far as possible.

Guideline: REPORTING		
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IV.2. INVESTIGATION

Once reported to the parish health department, the Medical Officer (Health) is responsible for ensuring that an investigation form (Appendix 2) is completed for all reported cases.

The Medical Officer on the hospital ward should complete the investigation form for all cases (suspected or confirmed) within 2 weeks and submit them to the parish Medical Officer of Health. The Public Health Nurse assigned to the case investigation should ensure that this is done within the time specified.

Once the investigation form is received, the Medical Officer (Health) should read it to ensure completeness and assign a classification, e.g. confirmed or discarded, if possible. A copy of this investigation report should then be forwarded to the Surveillance Unit where the Medical Officer, Surveillance will verify the classification. If unable to classify the case at parish level or at the Surveillance Unit, then the investigation form should be forwarded to the Paediatric Consultant for comments and advice.

V. SECONDARY PREVENTION

For all individuals who have had an initial attack of Rheumatic Fever, whether or not they have Rheumatic Heart Disease, continuous administration of antibiotics is a must to prevent re-infection of the upper respiratory tract by Group A Streptococcus. This has been proven to reduce the risk of recurrence of Rheumatic Fever and worsening morbidity and mortality.

There are different regimens for secondary prophylaxis but the regular intra-muscular injection of Benzathine Benzylpenicillin is the most effective treatment available. It is given every 28 days (4 weeks) but recent data indicates that if given every three weeks instead, it is even more effective in preventing recurrences of Rheumatic Fever, especially in high-risk patients such as adolescents or those with Rheumatic Heart Disease.

The injection should be given deep into the muscle as recommended so as to ensure maximum absorption and high serum levels. Care should be taken especially in adults, to ensure that the whole contents of the vial are mixed thoroughly, fully removed and injected slowly using a 20-gauge needle. The patient should be observed for at least 30 minutes to ensure no adverse effects.

Oral penicillin may also be used for secondary prophylaxis but the rate of recurrence of rheumatic fever is higher with this than with the benzathine benzylpenicillin. For patients who are allergic to penicillin or absolutely refuse to comply with the injections, oral sulfonamides or erythromycin is recommended. See Table 5.

Table 5: Recommended Antibiotics for Secondary Prophylaxis

Antibiotic	Route	Dose
Benzathine benzylpenicillin	IM	<30 kg: 600,000 IU every 3–4 weeks ≥30 kg: 1,200,000 IU every 3–4 weeks
Phenoxymethylpenicillin	Oral	250 mg 2 times daily
Sulfonamide (e.g. sulfadiazine, sulfadoxine or equivalent)	Oral	<30 kg: 500 mg daily ≥30 kg: 1.0 g daily
Erythromycin	Oral	250 mg 2 times daily

The duration of secondary prophylaxis should be adapted to the individual patient. Generally, patients without carditis in a previous attack should receive prophylaxis for a minimum of five (5) years after the last attack or at least until age 18 years whichever is longer. Those persons susceptible to recurrences for example persons working with children and health workers should continue prophylaxis until age 25 years. Patients with cardiac involvement in the initial attack but with evidence of healed carditis should continue prophylaxis at least until age 25 years and longer if

environmental conditions or other risk factors warrant it. Patients with chronic valvular rheumatic heart disease should receive prophylaxis for life unless taken off by a cardiologist.

Table 6: General Principles for Duration of Secondary Prophylaxis

Patient	Duration
No Carditis/No RHD	To 18 years and at least five years after the last attack
Documented Carditis – Healed (no evidence of Rheumatic Heart Disease)	At least to 25 years and often longer if warranted
Chronic Rheumatic Heart Disease	For Life
With Artificial Valves	For Life

35 yrs

Secondary prophylaxis should be continued during pregnancy. However, sulfonamides present a risk to the foetus and Penicillin or Erythromycin should be substituted. Special efforts should be made to ensure compliance especially among adolescents, as the risk of recurrence is greatest during this period.

Secondary prophylaxis should preferably be given from the Health Centre nearest to the patient's home, school or work. If the Health Centre is inconvenient, then the nearest Hospital should be the selected site. To avoid prophylaxis becoming due on a weekend, give return appointments of less than 28 days if necessary in order to bring the due dates back to a weekday. **Patients on oral medication should be monitored for compliance on a monthly basis.** Give a prescription for one month's supply then check compliance by counting remaining tablets.

Patients should be given a card on which their prophylaxis is recorded monthly. All patients should have a medical examination at least once per year by the Medical Officer in the clinic to check for Rheumatic Heart Disease. Any intervening occurrence of sore throats should be managed immediately. If there is any doubt about the physical examination, then the patient should be referred to the Internist in the region or the Cardiologist at the University Hospital.

VI. MANAGEMENT OF ANAPHYLAXIS

Anaphylactic reactions are an unavoidable aspect of the practice of medicine. Parenteral administration of medication usually produces more severe reactions than oral administration. If medication is given parenterally, the patient should remain under observation for 20– 30 minutes after administration.

A thorough history for drug allergy should be taken in every patient and all drugs should be properly labeled. Patients subject to anaphylaxis should be encouraged to purchase and wear a "Medic Alert" bracelet or necklace and should be advised to keep an identification card in their wallet or purse.

All health care workers should be trained in the diagnosis and management of anaphylaxis. The management should be rehearsed on a regular basis, at least twice per year. All health facilities should have an emergency tray with equipment and medication for the management of anaphylaxis at the point where injections are given (**see Table 7**). The parishes and regions must ensure that the minimum requirements are in place.

Table 7: Equipment and Medication for Management of Anaphylaxis

PRIMARY	• **Tourniquet** • **1-ml and 5-ml disposable syringes** • **Oxygen tank and mask/nasal cannula** • **Ambu-bag, oral airways,** laryngoscope, endotracheal tubes • **Large bore intravenous catheters and giving sets** • **Intravenous fluids (crystalloids and colloids)** • Aerosol beta 2 bronchodilators • Glucagon • **Normal saline / Hartmann's solution** • **Epinephrine (adrenaline) solutions (aqueous) 1:1000 and 1:10,000** • **Injectable Diphenhydramine or Phenergan** • **Injectable Corticosteroids (Hydrocortisone)** • Injectable Ranitidine or Cimetidine • Electrocardiogram

NB: Items in bold are the minimum requirements.

Table 7: Equipment and Medication for Management of Anaphylaxis (cont'd)

SUPPORTING	• Suction Apparatus • Sodium Bicarbonate • Aminophylline • Atropine • Nonlatex gloves • IV "set up" with needles, tubing and tape
OPTIONAL	• Defribillator • Calcium Gluconate • Neuroleptics for seizures • Lidocaine

The signs and symptoms of Anaphylaxis are:

Early – Sensations of warmth, itching, especially in the Axillae and Groins

Feelings of anxiety or panic.

Progressive – Erythematous or urticarial rash

Oedema of face, neck or soft tissues

Severe – Bronchospasm (wheezing)

Laryngeal oedema (dyspnoea, stridor, aphonia, drooling)

Hypotension (shock), Arrhythmias, Cardiac Arrest.

The initial step in the management of anaphylaxis is a rapid assessment of the patient's status with emphasis on evaluation of the airway and the state of consciousness. If the airway is compromised, it should be secured at that time. Blood pressure and pulse should be obtained. The patient should be placed in the supine position with the feet elevated. This position may have to be modified if the patient is wheezing. Oxygen should be started and the patient's weight assessed to guide the dosage of medications.

The following is a more detailed guide for the management of Anaphylaxis:

1. Stop administration of the causal agent.

2. Establish airway (oral airway or endotracheal intubation if required).

10-15 L/minute

3. Administer oxygen 100% (6 L/minute).

4. Assist ventilation if required (Bag and Mas

5. Place the patient in a recumbent position
where injection was given in the thigh).

6. Administer Adrenaline 1:1000 (1 mg/ml) intramuscularly.

Adults – 0.3–0.5 ml

Children – 0.01 ml/kg body weight

This may be repeated at 5-minute intervals.

7. Establish intravenous line and expand blood volume rapidly with crystalloid or colloid solution. For adults 500 ml rapidly then slowly there after. For children 20 - 30 ml/kg in the first hour.

8. Administer intravenous adrenaline only if:

repeatedly

(a) there is no response to intramuscular adrenaline; or

(b) there is circulatory failure and shock.

The dose is:

1 ml in 10 ml of Saline

- **Adults** – either adrenaline 1:10,000 (0.1 mg/ml) 5 ml administered *slowly over ten minutes* or adrenaline 1:1000 (1.0 mg/ml) 0.5 ml diluted in 10 ml of normal saline and administered *slowly over ten minutes.*
- **Children** – either adrenaline 1:10,000 (0.1 mg/ml) 0.1 ml/kg administered *slowly over ten minutes* or adrenaline 1:1000 (1.0 mg/ml) 0.01 ml/kg diluted in 10 ml normal saline and administered *slowly over ten minutes.*

ADDITIONAL MEASURES

(a) Administer either intravenous Corticosteroid e.g. Hydrocortisone 2–6 mg/kg or Dexamethasone 0.1–0.3 mg/kg.

(b) Administer either intravenous Antihistamine, e.g. Promethazine 0.5–1 mg/kg or Diphenhydramine 0.5 - 1 mg/kg.

(c) If there is bronchospasm, administer either aerosol bronchodilator (salbutamol) or nebulized bronchodilator via face mask and/or intravenous aminophylline: 6 mg/kg over 10 minutes followed by infusion 0.6 mg/kg/hour.

SUPPORTIVE TREATMENT

1. Monitor vital signs frequently every 10 minutes and maintain patient under medical observation for at least 4 hours.

2. Give reassurance. Keep patient rested and avoid heat.

The mainstay of the treatment of anaphylactoid reactions is adrenaline and fluids. Adrenaline should be used early at the first suspicion of Anaphylaxis. It is safe and effective. Additional vasopressor drugs, e.g. Dopamine are rarely indicated and can only be used in an ICU setting. Patients on Beta Adrenergic Blockers will require larger volumes of fluid and atropine or glucagon to improve cardiac output.

Patients with severe Anaphylaxis should be referred to hospital once stabilized and require observation for 48 hours or more.

VII. MANAGEMENT OF DEFAULTERS

All cases of confirmed rheumatic fever and rheumatic heart disease should be placed in a register at the parish health department and at the selected site for secondary prophylaxis. Contact information including a locating address and next of kin should be included in the registers.

If a patient misses his/her appointment for secondary prophylaxis, attempts should be made to find the patient within two weeks and return him/her for treatment. The defaulted patient should be assigned to a community health aide whose responsibility it now becomes to locate the patient and bring him/her in for treatment. Attempts to locate the patient may need to be done outside of normal working hours. If unable to locate the patient, information should be sought from relatives or neighbours regarding the whereabouts of the patient. If he/she has moved to another parish, the information should be communicated to the parish Medical Officer (Health) of that parish and ask to have the patient located. At least three (3) attempts should be made to locate defaulted patients, commencing within two (2) weeks of the missed prophylaxis. These should be documented in the patient's records. If a patient has defaulted and not presented at least once in a consecutive two-year period, he should be removed from the register and placed on a list of "Inactive Rheumatic Fever Patients".

Home visits may be necessary to ensure patient adherence to secondary prophylaxis.

VIII. PREVENTION OF INFECTIVE ENDOCARDITIS

Infective Endocarditis is a major threat to the patient with Rheumatic valvular heart disease. The patients and their relatives should be educated about this and the need for prevention with special antibiotic prophylaxis for certain procedures. They must be encouraged to inform their physicians and dentists and ensure compliance with the prophylaxis prior to invasive surgical procedures involving infected or contaminated tissue. Also for dental procedures in which bleeding may occur.

The recommended prophylaxis for **dental, oral or respiratory tract procedures** under local anaesthetic for standard-risk patients is:

- **For adults** – Amoxycillin 2 grams or Phenoxymethyl Penicillin 2 grams, orally, 1 hour before the procedure.

- **For children** – 50 mg/kg orally 1 hour before procedure.

If allergic to Penicillin, recently received Penicillin in the past 2 weeks, or on long-term Penicillin:

- **For adults** – Erythromycin, 1.5 grams, orally 1 hour before the procedure or Azithromycin or Clarithromycin 500 mg orally 1 hour before the procedure.

- **For children** – Azithromycin or Clarithromycin 15 mg/kg orally 1 hour before the procedure.

Clindamycin 600 mg (adults) or 20 mg/kg (children) may also be given orally 1 hour before the procedure.

All may be given IV or IM within 30 minutes of the procedure if unable to take oral medications.

For gastrointestinal and genitourinary tract procedures, the recommended prophylaxis is:

- **For adults** – Ampicillin, 2 grams, IM or IV, plus Gentamicin, 1.5 mg/kg (max 120 mg), intravenously, within 30 minutes of starting procedure and Ampicillin, 1 gram, IM or IV or amoxicillin 1 gram orally 6 hours later.

- **For children** – Ampicillin 50 mg/kg (max 2 grams) IM or IV plus Gentamicin 1.5 mg/kg within 30 minutes of starting the procedure followed by Ampicillin 25 mg/kg IM or IV or Amoxicillin 25 mg/kg orally 6 hours later.

If allergic to Penicillin:

- Vancomycin 1gram (adult) or 20 mg/kg (children) IV over 1–2 hours should be substituted for Ampicillin. The infusion should be completed within 30 minutes of starting the procedure.

Highest risk
Prosthetic cardiac valve
Previous IE
Congenital heart disease

IX. RECORD KEEPING – REGISTERS AND DATABASES

All confirmed rheumatic fever and rheumatic heart disease patients should be put on a register kept at the health centre level or site of prophylaxis. These should be compiled into a parish register kept at the parish health department. The register should be a line listing of the cases detailing their name, date of birth, sex, locating address, next of kin and the presence or absence of carditis. This register at parish level could be in the format for secondary prophylaxis or kept in a quire book. The record of secondary prophylaxis at health facilities should be a separate register and not be used for the parish register. The parish register should include all cases of rheumatic fever ever reported with no removal of names but indications of death or cessation of prophylaxis noted. The prophylaxis register should include those cases on prophylaxis for the year.

The clinic prophylaxis registers should be updated on a monthly basis if there are new cases, migrations or deaths. If cases are being removed, a report on each should be submitted to the coordinator. The registers should be monitored weekly/biweekly for defaulters and efforts made to find them as outlined previously.

The electronic database should be used to enter confirmed patients' information based on the investigation form completed. This database may be used as the register at parish level. The cases on register and their record of secondary prophylaxis may be used to calculate incidence and prevalence of rheumatic fever as well as compliance with secondary prophylaxis. Parishes may use this data to monitor and improve programme management.

The national database will be kept at the Surveillance Unit. All investigation forms should be submitted to the Unit so that confirmed cases may be included on this database. Monthly reports and details of patient secondary prophylaxis record should also be submitted in hard copy. The national database will be used to prepare annual programme reports including incidence, prevalence and secondary prophylaxis compliance. The weekly hospital active surveillance reports should be used to monitor possible additions to this register and the relevant parishes followed up for the investigation reports.

Guideline: RECORD KEEPING – RE GISTERS AND DATABASES		
Date Revised:	Distribution to hospitals and health centres II to V	Index: IX
Approved by: Director, Family Health Services		

X. REMOVAL OF PATIENTS FROM THE PROPHYLAXIS REGISTER

The following are the criteria for removal of patients from the Rheumatic fever register:

- Patients who have died or migrated out of the country should be removed from the register.

- Patients who meet the criteria for cessation of secondary prophylaxis may be removed from the register. However such patients must be examined by a doctor to ensure that there are no signs/symptoms of current rheumatic fever or clinical evidence of rheumatic heart disease (murmur).

- Defaulters who have not attended even once in the preceding two years may be removed from the register.

- Defaulters who meet the criteria for cessation of secondary prophylaxis may be removed from the register (even if not located for the purposes of a medical examination).

Once the decision is made regarding removal from the register, the reason(s) should be documented and the patient taken off the register immediately.

Defaulters or patients removed from the prophylaxis register should be placed on a list of "Inactive Rheumatic Fever Patients". They should be re-instated on the register if they are found and don't meet the criteria for cessation of secondary prophylaxis.

All cases removed from the register should be re-instated if they are once again diagnosed as having Rheumatic Fever.

XI. THE ROLE OF THE DOCTOR AND NURSE

All members of the health team have a role to play in the prevention and management of rheumatic fever. However the doctor and the nurse have specific duties to perform as follows:

Responsibility of the Medical Officer (Health), District Medical Officer and Medical Officer in the clinic or Accident and Emergency Department

1. Be responsible for the primary prevention of RF/RHD and ensure that "strep" throats are adequately treated.

2. Ensure that patients with suspected acute rheumatic fever are referred immediately to hospital for management and that the parish health department and surveillance unit are notified.

3. Know how many patients are registered for prophylaxis and ensure maintenance of the register.

4. Be aware of the secondary prophylaxis coverage rate and patient compliance rate in the area at all times.

5. Ensure that efforts are made to contact defaulters within two (2) weeks of a missed appointment.

6. Review the programme with the nurse-in-charge of the programme on a monthly basis.

7. Review and sign the monthly reports before submission to the parish coordinator.

8. Ensure that all RF/RHD patients are seen at least annually for a physical examination and that patients with RHD receive appropriate cardiology consultation and follow up.

9. Decide when prophylaxis may be discontinued, according to the guidelines and complete a report for submission to the parish coordinator.

10. Know the steps to be taken in the event of anaphylaxis and ensure rehearsal of these regularly with other members of staff.

Responsibility of the Public Health Nurse, Nurse Practitioner and Registered Nurse in the Health Centre and the Accident and Emergency Department

1. Investigate all reported suspected cases to ensure that the diagnosis is confirmed.

2. Administer the benzathine benzylpenicillin injections as per the guidelines.
3. Keep a register of RF/RHD patients treated in the health facility.

4. Keep a record of benzathine benzylpenicillin injections given to the patients on the forms provided.

5. Ensure the patient prophylaxis cards are kept up to date.

6. Ensure that all patients and/or their parents or guardians are educated about the disease and understand the need for secondary prophylaxis.

7. Ensure that defaulters are contacted and encouraged to attend for prophylaxis within two (2) weeks of a missed appointment.

8. Submit a report on each patient who has been removed from the register to the parish coordinator.

9. Ensure that programme reports are submitted to the parish coordinator on a monthly basis using the forms provided.

10. Ensure that adequate supplies of drugs, materials and supplies, especially penicillin, adrenaline, fluids, needles and syringes are available and easily accessible for patient prophylaxis and management of anaphylaxis.

11. Know the steps to be taken in the event of anaphylaxis and rehearse them with other members of staff.

XII. MONTHLY PROGRAMME REPORTING

Each parish should submit a monthly report on the programme, to the Surveillance Unit, using the form provided (see Appendix 3). The report should be submitted by the 15th of the following month. This report should be a summary of the reports from all prophylaxis sites in the parish. This includes the hospitals if they are used for prophylaxis. All areas should be completed as best as possible. Zeros should be used instead of leaving the space blank. The report should be accompanied by a written explanation for cases removed from the register.

On a quarterly basis, copies of the parish registers must be submitted to the surveillance unit for updating of the national register.

A compliant patient is one who has received at least 90% (12 of 13 doses) of the benzathine benzylpenicillin injections per year or has taken 90% of the oral medication prescribed for prophylaxis. Centre compliance is calculated by dividing the total number of compliant patients by the total number of patients registered. The centre prophylaxis coverage is calculated by dividing the total number of injections given by the total number of injections due. However, efforts should be made to ensure that secondary prophylaxis is received on time.

The report should be reviewed by the parish Medical Officer (Health), commented on and signed prior to submission to the Surveillance Unit.

Guideline: MONTHLY PROGRAMME REPORTING		
Date Revised:	Distribution to hospitals and health centres types II to V	Index: XII
Approved by: Director, Family Health Services		

XIII. HEALTH PROMOTION AND EDUCATION

The main goal of health promotion and education is to increase awareness on primary prevention as well as adherence to secondary prophylaxis. All members of the health team are responsible for various aspects of health promotion and education and should have in-service sessions to update their knowledge and skills. Rheumatic fever education should be integrated into other health promotion and education activities and strategies. The main target audiences should be children in schools, teachers and parents/guardians. Promotional and educational messages should be disseminated through workshops, training sessions and printed material. The electronic media may also be used. Rheumatic fever patients should also receive education at each visit for prophylaxis and adherence emphasized.

XIV. APPENDICES

1. Class I Reporting Form – Individual Notification
2. Investigation Form
3. Control Programme (Monthly Report)

CLASS I REPORTING FORM – INDIVIDUAL NOTIFICATION

Date of Report: _____ / ____ /_____ (DD/MM/YY) NEW CASE / UPDATE on previously reported case (Circle One)

Diagnosis: :

Case Demographic Information

Name (including pet name): _____ Sex: ____ Age:_____ D.O.B ____ / ____ / _____(dd/mm/yy)

Address (Including Landmark) _____

_____ Parish: _____

Workplace/School _____ Occupation: _____

Home Phone No.: _____ Wk Phone #: _____ History of overseas travel in past 4-6 weeks? Y / N
Specify area/country:

Name of NOK/Parent: _____ Relationship to case: _____

Address of NOK/Parent: _____ Phone No.: _____

Clinical Information:

Symptoms: _____

_____

Date of onset: ___ / ___ / _____ (dd/mm/yy) Date seen: ___ / ___ / _____ (dd/mm/yy)

Specimens Taken? Y / N Type: _____

Specimen Date: ___ / ___ / _____ (dd/mm/yy) Laboratory:_____

Hosp./Facility Name: _____

Medical Record #: _____

Case admitted to Hosp?: Y / N (Circle one)

Date of Admission: ____ / ____ / _____ (dd/mm/yy)

If dead, Date of Death: ____ / ____ / _____ (dd/mm/yy)

Notifier information

Name of notifier: _____ Phone #: _____

Address: _____ Email address: _____

Comments:_____

_____

Ministry of Health, Surveillance Unit. November, 2000

Appendix 1

Appendix 2

MINISTRY OF HEALTH, JAMAICA
RHEUMATIC FEVER / RHEUMATIC HEART DISEASE CONTROL PROGRAMME

INVESTIGATION FORM

Case Identification

Notification Site: _____ Parish: _____

Name of patient: _____ Gender: ☐ Male ☐ Female

Address: _____ Date of Birth: ____ / ____ / _____ (dd/mm/yyyy)

_____ Name of parent/guardian: _____

_____

School Attended: _____ Telephone No.: _____

Clinical Information

Major manifestations	Yes	No	Unk	Minor manifestations	Yes	No	Unk.
Carditis	☐	☐	☐	Joint pain	☐	☐	☐
Polyarthritis	☐	☐	☐	High temperature (fever)	☐	☐	☐
Chorea	☐	☐	☐	History of rheumatic fever	☐	☐	☐
Erythema marginatum	☐	☐	☐	Previous rheumatic heart disease (RHD)	☐	☐	☐
Subcutaneous nodules	☐	☐	☐	Positive C-reactive protein test	☐	☐	☐
				Increase erythrocyte sedimentation rate (ESR)	☐	☐	☐
				Increase in the PR interval (ECG)	☐	☐	☐

Laboratory evidence of recent Streptococcal infection

	Yes	No	Unk
Elevated ASTO	☐	☐	☐
Throat swab positive for GpAβ-haemolytic Strep.	☐	☐	☐

Initial attack: ☐ Yes / ☐ No / ☐ Unknown

If No, year of initial attack: _____ (yyyy)

Number of recurrences since initial attack: _____

Prophylaxis received in previous year: ☐ Yes / ☐ No

No. of Benzathine Penicillin injections received: _____

Oral prophylaxis type: _____ Regular ☐ / Irregular ☐

Cardiac Involvement

	Suspected	Positive	Negative
Mitral Stenosis	☐	☐	☐
Mitral insufficiency	☐	☐	☐
Aortic stenosis	☐	☐	☐
Aortic insufficiency	☐	☐	☐
Organic lesion of the tricuspid valve	☐	☐	☐
Heart Failure	☐	☐	☐
Bacterial Endocarditis	☐	☐	☐

Severity of heart damage: ☐ None / ☐ Very little / ☐ Moderate / ☐ Serious / ☐ Not Determined

Echocardiogram Findings _____

_____

_____

_____

Hospital Information:

Hospitalized: ☐ Yes / ☐ No

If Yes, name of Hospital : _____

_____

Date of Admission: ___ /___ /____(dd/mm/yyyy)

Docket No.: _____

Date of Discharge: ___ / ___ /____(dd/mm/yyyy)

Duration of stay: _____ (No. of days)

Classification

☐ Suspected ☐ Discarded ☐ Confirmed

Investigator's Comments:

_____

_____

_____

Name: _____

Position: _____

Signature: _____

Date of Investigation: _____ / _____ / _____

M.O. (H) Comments: _____

_____

_____

M.O.(H) Signature: _____

Date: _____ / _____ / _____

PLEASE FAX INVESTIGATION FORMS TO THE SURVEILLANCE UNIT AT 967-1280

Appendix 3

MINISTRY OF HEALTH, JAMAICA
RHEUMATIC FEVER / RHEUMATIC HEART DISEASE
CONTROL PROGRAMME (MONTHLY REPORT)

TO: Surveillance (Epi) Unit
FAX: 967-1280
MONTH: _____

FROM: PARISH: _____
FAX: _____
DATE: _____

REGISTRATION OF CASES

TOTAL NO. OF CASES ON REGISTER _____
NO. OF OLD CASES _____
NO. OF NEW CASES _____
NO. OF CASES REMOVED FROM REGISTER _____

NAMES OF CASES TRANSFERRED INTO PARISH — REASONS:

_____ _____
_____ _____
_____ _____
_____ _____

NAMES OF CASES TRANSFERRED **OUT** OF PARISH — REASONS:

_____ _____
_____ _____
_____ _____
_____ _____

NO. OF DEATHS _____

NAME OF DEAD CLIENT — CAUSE OF DEATH:

_____ _____

PROPHYLACTIC COVERAGE

NO. OF INJECTIONS DUE FOR MONTH _____
NO. OF INJECTIONS GIVEN FOR MONTH _____
NO. OF REGISTERED CLIENTS FULLY COMPLIANT _____
NO. OF CASES OF ANAPHYLAXIS _____

HOSPITAL ADMISSIONS

NO. OF SUSPECTED CASES _____
NO. OF CONFIRMED CASES _____
NO. OF INITIAL ATTACKS _____
NO. OF RECURRENT CASES _____
NO. OF CASES WITH CARDITIS _____

PRIMARY PREVENTION

NO. OF HEALTH EDUCATION SESSIONS HELD:

		TOTAL AUDIENCE
IN CLINICS	_____	_____
IN SCHOOLS	_____	_____
IN COMMUNITIES	_____	_____
OTHER	_____	_____

NO. OF EDUCATIONAL MATERIALS DISTRIBUTED/DISPLAYED _____

NO. OF HEALTH WORKERS TRAINED:

DOCTORS	_____
NURSES	_____
HEALTH AUXILIARIES	_____

Name of Co-ordinator: _____ Signature: _____ Date: _____

Comments: _____
_____

M.O.(H). Comments: _____
_____

M.O.(H) Signature: _____ Date: _____

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