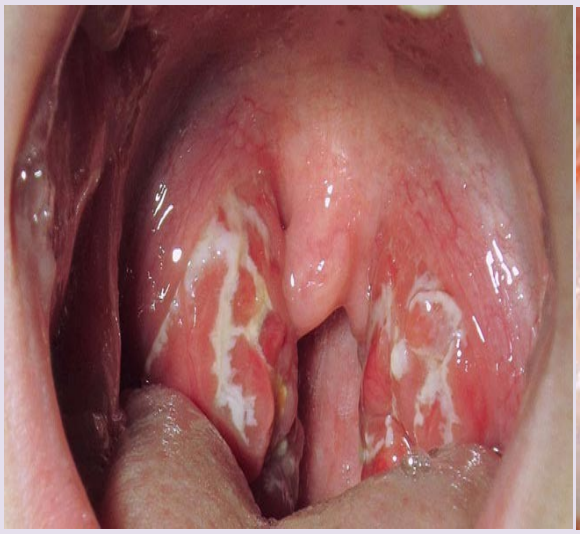
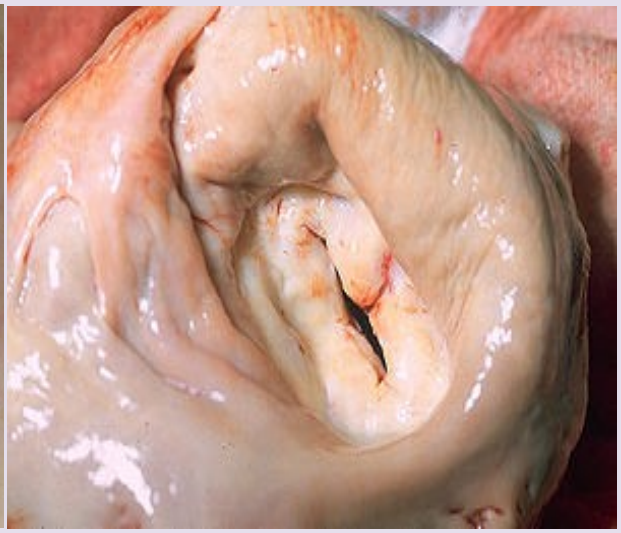
Federal Democratic Republic of Ethiopia

Ministry of Health





NATIONAL TRAINING ON RHEUMATIC HEART DISEASE PREVENTION & CONTROL FOR HEALTH CARE WORKERS IN ETHIOPIA:

Participant's Manual

September 2017

Addis Ababa

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ABBREVIATIONS & ACRONYMS

ARF:	Acute Rheumatic fever
ACEI:	Angiotensin converting enzyme inhibitor
BP	Blood pressure
RHD	Rheumatic Heart Disease
CT	Computerized Tomography
ECG	Electrocardiogram
Echo	Echocardiography
ESR	Erythrocyte sedimentation rate
GABHS	Group A β hemolytic streptococci
HF/CHF	Heart failure
IE	Infective endocarditis
MRI	Magnetic resonance Imaging
PAD	Peripheral artery disease
PR	Pulse Rate

ACKNOWLEDGMENTS

The Federal Ministry of Health would like to extend its sincere appreciation to all those who contributed to the development of this training package. It is not possible to mention all who participated in the preparation of this document; however, we would like to acknowledge the following professionals for developing the training materials:

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The following professionals provided valuable feedbacks and were involved as editors to the ARF/RHD training manual:

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Dr Abraha Hailu	Mekelle University	Internist, Consultant Cardiologist
Dr Araya Giday	Hawassa University	Internist, Consultant Cardiologist

Additionally the Ministry would like to thank Friends of Children Heart Fund –Ethiopia for supporting this initiative which will help us address this deadly but silent disease afflicting significant proportion of our young population.

It is hoped that this training material and tools will help fill the information and skill gap of our health care workers so that they will deliver high quality uninterrupted care to patients with sore throat, ARF/RHD.

Biruck Kebede (BSc, MPH)

A/Director, Diseases Prevention and Control Directorate

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INTRODUCTION AND RATIONALE ABOUT RHD TRAINING

Ethiopia is one of the developing countries with high burden of communicable diseases. Rheumatic fever follows bacterial pharyngitis and ends up with rheumatic heart disease as long term sequel. The prevalence of rheumatic heart disease is around 1.4% in 6-18 years old individuals based on echocardiographic school surveys. Rheumatic heart disease is the main cardiovascular disease in Sub-Saharan African countries including Ethiopia.

Increasing burden of non-communicable is being seen in recent years due to urbanization and change in life styles. Less attention was being given to these non-communicable disease due to lack of infrastructure and overburden by communicable diseases and hence there have been no national NCD treatment guideline and training materials for health workers in Ethiopia.

In recent years FMOH has developed strategic plan to control these diseases. The ministry has undertaken STEPS survey to estimate the burden of these diseases and recently has developed Clinical and Programmatic Management Guidelines on Major NCDs.

This manual is prepared as part of NCD control activities and intends to be used as a training manual for primary health care workers practicing at health center and primary hospital levels. It can also be a useful additional training for hospital staff working with child health and general medicine.

This RHD training manual has 7 units covering Sore throat, Rheumatic fever, Rheumatic Heart disease, RHD control program and Monitoring evaluation of RHD Programs.

The manual focuses on early identification and treatment of bacterial pharyngitis, rheumatic fever and rheumatic heart disease at health center and primary hospital where majority of patients can easily get access and get treated.

RATIONALE OF THIS TRAINING

Baseline assessment prior to the design of this course showed that the service being given at hospital and health center level lacks depth and quality. The service is almost nonexistent at health center level. This training manual is therefore developed to improve the quality of care at hospital level, strengthen referral systems between hospitals and health centers and strengthen NCD services at health center level.

TRAINING OBJECTIVES

Major Objective:

- After completing this course, health care professionals in Ethiopia will be able to, understand, diagnosis, manage and prevent ARF/RHD

Enabling Objectives:

To meet the course objective, learners will be able to:

1. Understand Tonsilo-pharyngitis
2. Differentiate between bacterial and viral tonsillo-pharyngitis
3. Differentiate between tonsillo-pharyngitis and tonsillar hypertrophy
4. Understand the pathogenesis of Acute Rheumatic Fever
5. List manifestations of acute Rheumatic Fever
6. Discuss the criteria to diagnose Acute Rheumatic Fever
7. Identify patients with ARF and Refer
8. Follow patients with ARF
9. Identify and refer patients with Rheumatic Heart Disease
10. List complications of RHD
11. Follow patients with RHD.

TARGET AUDIENCE

The main target audiences are B. Sc nurses, Health officers and GPs working at HC and Hospital level. Parts of the material can also be used to sensitize or train health managers at all levels and community health workers. The national guideline accompanying this module has already outlined what these cadres can do at primary level and how the secondary and tertiary levels may be utilized appropriately through referral and linkages. The training package hence focuses on what can be done at health center level, hospital levels.

CORE COMPETENCIES

The trainees should demonstrate the following core competencies after successful completion of the training.

1. Application of the concepts and principles of Sore throat, ARF and RHD management
2. Taking and documenting patient history and do basic physical examination
3. Assessing and managing patients with tonsilopharyngitis, ARF and RHD according to the national ARF/rheumatic heart disease management guideline
4. Acquiring knowledge, skills and confidence in the diagnosis of tonsilopharyngitis and use of medications to manage bacterial pharyngitis to prevent rheumatic fever and other medications to manage complications of RHD
5. Communicating with patients and their families and providing ethical, patient-centered care
6. Recognizing the importance of working as team for effective ARF/RHD management
7. Assessing and managing the psychosocial, spiritual and cultural dimensions of through counseling and other non-pharmacological approaches
8. Advocate for Rheumatic Fever/Rheumatic Heart Disease prevention

COMPONENTS OF ARF/RHD TRAINING PACKAGE

This ARF/RHD course is built around use of the following components:

- **A participant's manual** containing need-to-know information on ARF/RHD and also containing questionnaires, exercises and checklists, which break down the skill or activity into its essential steps.
- **A Facilitators guide**, which includes questions, answers keys and detailed information for conducting the course.
- **PowerPoint presentations** which are explained by figures an numbers with foot notes.
- Well-designed teaching aids and other training aids
- National Guidelines on Clinical and Programmatic Management of Major NCDs.

CONDUCT OF THE TRAINING

This course will be facilitated by health care professional who took the national training of trainers' course. They TOTs should be clinicians with basic training on internal medicine or pediatrics. They can be cardiologists, internists, pediatricians or general practitioners. Experienced health officers and BSC nurses could also serve as TOTs with initial support from specialists.

In keeping with the training philosophy on which this course is based, all training activities will be conducted in an interactive, participatory manner. To accomplish this requires that the role of the trainer continually change throughout the course. For example, the trainer is an instructor when presenting a classroom demonstration; a facilitator when conducting small group discussions or using role plays; and shifts to the role of coach when helping participants practice a procedure. Finally, when objectively assessing performance, the trainer serves as an evaluator.

RHD TRAINING COURSE AGENDA

National Training on Prevention and Control Rheumatic Heart Disease

Time	Activity	Responsible person
	Day 1	
8:30-9:30	Registration Opening and overview welcome and Introduction Course goals and objective, Material and schedule pretest	
9:30-10:00	Unit 0: Introduction about Acute Rheumatic fever and RHD	10 slides
10:00-10:20	Tea Break	

10:20-12:30	Unit 1: Acute Tonsillopharyngitis Reading Case study Group discussion Presentation	28 slides
Lunch		
1:30 – 3:30	Unit 2: Acute Rheumatic Fever- Part I Reading Group discussion Presentation	43 Slides
3:30-4:00PM	Tea Break	
4:00-5:30PM	Unit 2: Acute Rheumatic Fever- Part II Reading Case study 1 and 2 Group discussion Presentation	
	Day 2	
8:30 -9:00 9:00- 10:20	Recap Unit 3: Secondary Prophylaxis to Prevent Recurrent RF Reading Group discussion Presentation	26 slides
10:20-10:40	Tea break	
10:40-12:30	Unit 4 - Diagnosis and Management of Rheumatic Heart disease Reading Group discussion Presentation	28 slides
Lunch		
1:30-3:30	Unit 5:Management of Complications of Rheumatic Heart Disease Part I Reading Case study 1and 2 Group discussion Presentation	60 slides
3:30-3:50	Tea Break	
3:50-5:30	Unit 5:Management of Complications of Rheumatic Heart Disease Part I Reading Case study Group discussion Presentation	
	Day 3	
8:30-9:00	Recap	
9:00 –10:30	Unit 6: RHD Prevention and Control Program Reading Discussion Presentation	27 slides
10:30-10:50	Tea break	
10:50-12:30	Unit 7: Monitoring and Evaluation of RHD Prevention and Control Program Reading Discussion	23 slides

	Presentation	
12:30- 1:30	Lunch	
1:30-3:00	Post test course evaluation certification closing ceremony	

UNIT 1: Diagnosis and Management of Tonsillopharyngitis

Learning Objectives

At the end this module the participants will be able to:-

- Describe the characteristics of Group A Beta Hemolytic Streptococci (GABHS)
- Describe the epidemiology of GABHS tonsillopharyngitis
- Explain the difference between viral and GABHS tonsillopharyngitis
- Outline the diagnostic modality of GABHS tonsillo-pharyngitis
- Outline the management of GABHS tonsillopharyngitis
- Explain methods of prevention of tonsillopharyngitis

1.1 Introduction.

Rheumatic fever is an inflammatory disease involving the joints, skin, heart and brain, which develops following an untreated or partially treated group A Beta-hemolytic streptococcal (GABHS) infection of the throat (streptococcal pharyngitis).
Up to 30% of sore throats in children and young people are caused by GABHS, and 0.3% to 3% of these young people with an untreated GABHS sore throat will develop RF.
After recovery from the initial episode of RF, up to 60% to 65% of patients develop valvular heart disease and the risk of RF recurrence following GABHS infection rises to 50%.
Identification and treatment of bacterial sore throat is an important component of Rheumatic Fever/Rheumatic Heart Disease Prevention and Control Program.

1.2 Etiology and Pathogenesis

Sore throat (Tonsillopharyngitis) is a symptom caused by inflammation of pharynx, tonsils or other surrounding structures. The inflammation can be due to infectious causes (bacterial or viral) or non-infectious causes related to environmental exposure such as air pollution and allergens.

Viruses are the predominate causes of sore throats, however Group A Beta hemolytic Streptococci are the commonest bacterial causes of sore throat(20-40% of sore throats in children and 5-15% of sore throats in adults).
GAβHS are Gram-positive, non-motile coccoid shaped bacteria that tend to grow in pairs or chains (see figure 1 below). They are characterized by local invasion and release of extracellular toxins (Streptolysins S and O) and proteases. M protein fragments of certain serotypes of GAβHS are similar

to myocardial sarcolemma antigens and this similarity is the basis for subsequent occurrence of carditis as a component of rheumatic fever.

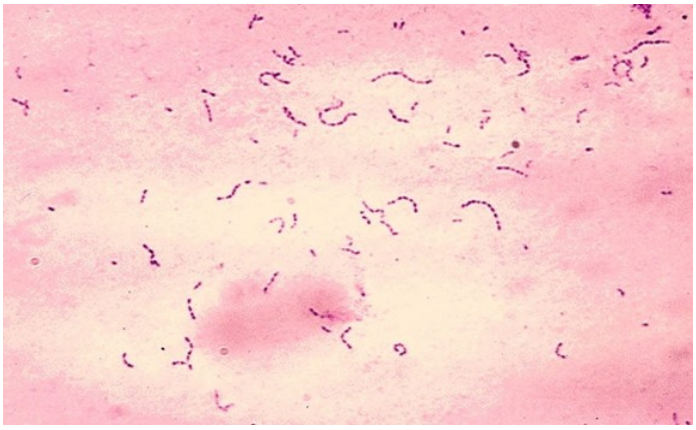


Figure 1.1 Group A Beta hemolytic Streptococci (GAβHS) seen in pairs and chains.

This organism may cause suppurative disease, such as pharyngitis, impetigo, cellulitis, myositis, pneumonia, and puerperal sepsis. It also may be associated with nonsuppurative disease, such as rheumatic fever and Acute Poststreptococcal Glomerulonephritis.

1.3 Epidemiology

Human beings are the natural reservoir for GABHS and exposed to these bacteria in the environment. Mostly they are spread through droplets of salivary or nasal secretions. Overcrowding, poverty, and close contact with person with streptococcal sore throat are considered risk factors for transmission Siblings, schoolmates and close contacts are at high risk. GAβHS has a potential to cause out breaks of diseases in the day care settings. The incidence of GAβHS pharyngeal infection is highest in children 5-15 years of age.

The incubation period for GABHS infection is 2 to 5 days. Throat and skin are common sites of GABHS infection. GABHS infections usually resolve without treatment however if left uuntreated it can lead to acute rheumatic fever in some people. Antibiotic treatment decreases severity of symptoms and reduces the risk of transmission to others after 24 hrs of treatment. Treatment also decreases the risk of acute rheumatic fever. Studies show that ARF associated with GABHS pharyngitis can be prevented if treatment is commenced within 9 days of appearance of the first symptoms.

- ***Group A β hemolytic streptococcus is the commonest bacterial cause of tonsilopharyngitis.***
- ***GABHS Tonsilopharyngitis has to be treated with antibiotics to prevent Suppurative and nonsuppurative complications.***

1.4 Clinical Presentations of Sore Throat

There is broad overlap between the signs and symptoms of streptococcal and viral sore throat. The ability to identify streptococcal sore throat accurately on the basis of clinical grounds alone is generally challenging. Acute GAβHS sore throat has certain characteristic epidemiologic and clinical features (see table 1 below) that may help the health professionals in the decision either to initiate antibiotics or not.

GAβHS sore throat is primarily a disease of children 5-15 years of age. It is relatively uncommon before 3 years of age. In temperate climates it usually occurs in winter and early spring.

Patients with GAβHS pharyngitis commonly present with:-

- Sudden onset of sore throat or Pain on swallowing
- Fever above 38^0C
- Headache, nausea, vomiting and abdominal pain may be present especially in children.

And on examination patients have:-

- Tonsilopharyngeal erythema
- Enlarged tonsils with exudates
- beefy red swollen uvula, petechia on the palate and scarlantiform rash
- Tender and enlarged anterior cervical lymph nodes (lymphadenitis),see figure 2 below.

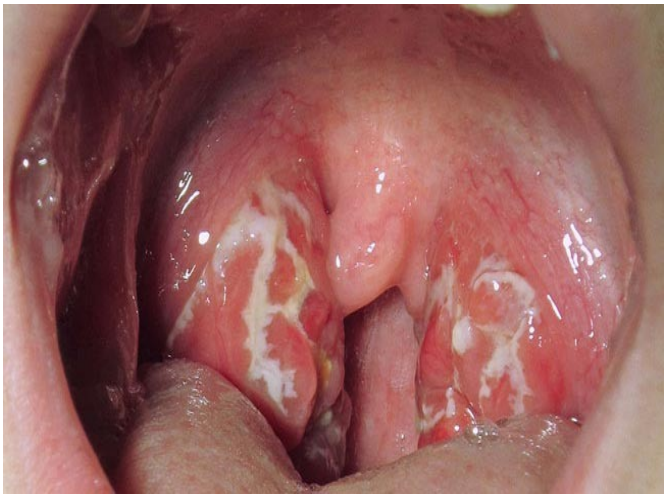


Figure 1.2. Tonsilar swelling with erythema and exudates.

Viral Tonsillopharungitis

Cough, runny nose conjunctivitis, hoarseness, coryza, anterior stomatitis, discrete intra-oral ulcerative lesions, viral exanthema, diarrhea and absence of fever strongly suggest a viral rather than a streptococcal etiology.

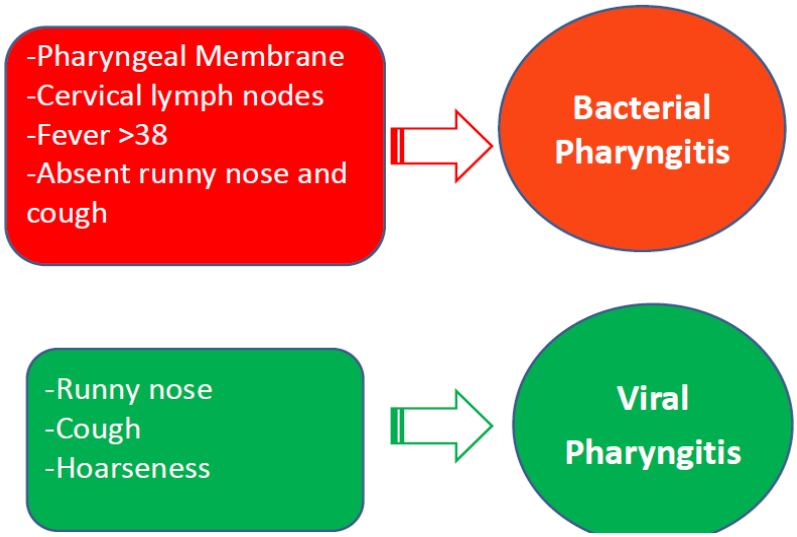


Figure 1. 3: Bacterial Vs Viral Pharyngitis

1.5 DIAGNOSIS OF BACTERIAL SORE THROAT

Etiologic diagnosis of sore throat is always a challenge even for experienced physicians. Yet it is extremely valuable to differentiate viral causes of sore throat from that caused by GAβHS to initiate the appropriate antibiotic therapy.

The diagnosis of streptococcal pharyngitis (GABHS) can either be clinical or using clinical criteria supported by laboratory investigations.

The gold standard diagnostic method is by using a Clinical Prediction rule (CPR) supported by rapid antigen test (RAT) and/or throat culture.

A. THROAT SWAB CULTURE

- Remains the "**gold standard**" for the diagnosis of GAβHS pharyngitis
- Not available in health centers and most hospitals
- Delay in getting results at least 48 hrs.
- Many people are asymptomatic carriers (10% of school age children) and can be culture positive without sore throat
- Technical errors (impact on the results)
- Cost of test is high

B. Rapid Antigen Detection Test for GABHS

- detects the group A carbohydrate of GAβHS
- used in office basis and result can be ready in 10 minutes
- Highly specific (>95%) but less Sensitive than culture
- Not validated in Ethiopia
- Expensive

C. Anti Streptolysin O (ASO) antibody Titer

- Titers increase only 7-14 days after the onset of infection and remain high for weeks.
- NO role in diagnosis of Acute Tonsillopharyngitis.

D. Clinical Decision Rules (CDR)

No single symptom or sign is useful when used alone but combination of signs and symptoms have been studied and have shown high sensitivity and reasonable specificity for diagnosis of bacterial tonsillopharyngitis in low income countries.

Table 1.1 : The Centor Clinical Decision Rule (CDR).

Symptoms or Signs	**Points**
History of high fever or (measured ≥ 38°C)	1
Absence of cough and rhinorrhoea	1
Tender anterior Cervical adenopathy	1
Tonsillar swelling or exudates	1

Interpretation of CDR

≥ 2 points treat as GAβHS pharyngitis (with antibiotic) ,

< 2 points, treat as viral pharyngitis (no antibiotic)

In Ethiopia it is recommended to use clinical decision rule for the diagnosis of bacterial tonsillopharyngitis.

1.6 Management of sore throat

Objectives of bacterial sore throat therapy

- to improve clinical symptoms and signs;
- for the prevention of suppurative complications (eg Peritonsilar abscess, cervical lymphadenitis, mastoiditis and possibly other invasive infections);
- for the reduction of transmission of GAβHS to family members, classmates and other close contacts of the patient;
- to allow for the rapid resumption of usual activities
- for the prevention of Acute Rheumatic Fever;

General Management

- Analgesics/Antipyretic (paracetamol) for relief of pain and fever.
- Identification and early treatment of complications
- Avoid cold drinks
- Hydration and rest may be needed.

Table 1.2: Specific (Antimicrobial therapy).

First Line	Alternative	If Patient is allergic to penicillin:
Benzathine penicillin G Dose: Wt. < 30kg: 600,000 IU IM stat. Wt. ≥ 30 kg: 1.2 million IU IM stat. **Follow Safe BPG Injection Procedures!**	**Amoxicillin** Dose **:** Children < 7years: 50 mg/kg per day in three divided doses for 10 days. Age ≥ 7Years: 500mg PO TID for 10 Days	**ERYTHROMYCIN** Dose: • Children < 7 years: 250 mg BD for 10 days • Age ≥ 7 years: 500 mg BD for 10 days

BPG is the drug of choice for GAβHS sore throat

Why Benzathine Penicillin is the drug of choice?

Benzathine Penicillin G is preferable to the other alternative PO antibiotics for treatment of tonsillopharyngitis.

- It is a single injection while oral treatment needs 10 whole days to be effective
- BPG has better bactericidal effect than oral antibiotics
- GAβHS are universally sensitive for penicillin
- Oral macrolides (Erythromycin and azithromycin) show clinical improvement but no eradication of organism.
- Cost effective, evidence based.
- Parents and patients more satisfied.

Is it cost-effective to administer BPG for all cases of suspected strep sore throat based clinical decision rule? Yes.

In patients with a sore throat and symptoms suggestive of a GABHS infection, antibiotic treatment using intramuscular BPG could reduce the risk of RF by up to 80%.

Tonsillectomy and Tonsillopharyngitis

Tonsillectomy is indicated if there are frequent episodes of tonsillitis or there is tonsillar hypertrophy with symptoms of upper respiratory tract obstruction.

Tonsillectomy may decrease but does not eliminate the occurrence of GABHS pharyngitis.

1.7 Prevention and Health Education

Families should be educated about:

1. The symptoms of GAS pharyngitis.
2. The serious consequences of untreated pharyngitis i.e. ARF and RHD and the need to consult medical personnel as early as possible to avoid complications.

3. The need to avoid pharyngitis by improving house ventilation and hygiene and avoid crowding.
4. Importance of adherence to a 10 days course of antibiotics in oral treatment.

Case study

W/O Almaz brought her five years old girl Helen to the outpatient clinic with acute onset of fever, severe throat, pain exacerbated by swallowing, headache and abdominal pain. No runny nose, no cough. On examination her temperature was 38.3^0C axillary, the tonsils were symmetrically enlarged, red with exudates. She had multiple enlarged painful anterior neck lymphadenopathies. No other abnormal findings detected.

Questions.

1. What is your clinical diagnosis of Helens' illness?
2. What tests do you need to reach at a diagnosis?
3. What is the most likely causative organism of her illness?
4. Which of the clinical presentation helps you to decide about the treatment you are going to give?
5. How would you like to treat Helen?
6. What is the drug of choice (type, dose and route of administration)?
7. What other additional Advice do you like to give to W/O Almaz?

Unit 1 Summary

- Group A beta hemolytic streptococcus is one of the commonest cause of bacterial tonsillopharyngitis.
- One of the non suppurative complications of untreated GABHS is Rheumatic fever
- Diagnosis of GABHS pharyngitis in national guideline is based on based on the Clinical Decision Rule (CDR): sore throat with exudate, fever > 38^oC, cervical adenopathy, no runny nose or cough .If Score $\geq$2 treatment with antibiotic is indicated.
- Drug of choice for GABHS is single injection of Benzathine Penicillin G IM .
    - Ask about penicillin Allergy.
    - Take measures to decrease the injection pain.
    - Emergency drugs such as epinephrine , suction and oxygen have to be available injection site
- Families have to be educated about the complications of GABHS pharyngitis and sequel of Rheumatic fever.

Unit 2: Acute Rheumatic Fever

Learning Objectives

At the end this module the participants will be able to:-

- Describe the epidemiology of ARF
- Explain the pathogenesis of ARF
- List the clinical features of ARF
- Clarify the diagnosis of ARF
- List the management of ARF
- Mention the prevention strategies of ARF

2.1 Introduction

ARF is a multisystem post infectious, non-suppurative sequelae of tonsillo-pharyngeal infection with Group A Beta hemolytic Streptococci (Streptococcus pyogenes).It usually affects children and young adults.RHD is the only long-term sequel of ARF or recurrent rheumatic fever. RHD manifests after several years (5-20 years) of ARF with heart failure or complications like atrial fibrillation, stroke or infective endcocarditis.ARF and RHD can easily be prevented by early identification and treatment of streptococcal pharyngeal infection. Currently, the 2015 AHA/ACC criteria are used to diagnose ARF.

2.2 Epidemiology

Burden of disease

RHD is a lifelong condition, which is often fatal if not treated properly.RHD is found all over the world, but most commonly affects women, adolescents and children living in conditions of poverty and overcrowding.

While the incidence and prevalence of ARF and RHD have been decreasing in developed nations since the early 1900s, it continued to be major cause of morbidity and mortality among children and young adults less than 40 years of age in developing nations. Rheumatic heart disease currently affects over 33 million people worldwide. RHD kills 275,000 people every year, even though it is a preventable disease. Ninety percent of the disease is confined to Sub-Saharan African and Sub Pacific Asia.

A few rich countries (including the USA and UK) and some Low and Middle Income Countries like Cuba have managed to reduce their burden of RHD, but other countries continue to struggle with the disease.

Reports from Africa indicated prevalence rates ranging from 2.4 to 14.5 as reported from Kinshasa, Kenya, and Egypt.

Recent school and community based studies in Ethiopia have shown the prevalence of RHD to be from 14-38/1000, in the age group 4-24 years, which is one of the highest in the world. Approximately 250, 000 people in the age group 5-15 suffer from RHD in Ethiopia and more than 500,000 people of all age groups live with RHD. Of these only few give history of Acute Rheumatic Fever. RHD is the major cardiovascular diagnosis accounting for 30-60% of all cardiac patients in registered in main hospitals of Ethiopia.

Patients usually come late with heart failure, stroke or during pregnancy with severe valvular disease Mortality from RHD may reach 12.5% every year in rural Ethiopia (which is much higher than global estimates of 1.2% annual mortality). It is also reported that 70% of RHD patients die before the age of 26 years.

Risk Factors for ARF

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

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

B. Age: ARF is most common in children between the ages of 5 and 15 years with a median age of 10 years. It is less common before 3 years of age and after the age of 35 years.But 20% of ARF cases are adults.

C. Sex:

- Rheumatic fever occurs in equal numbers in males and females, but the prognosis is worse for females than for males.

Table 2.1: Direct and indirect results of environmental and health-system determinants on ARF/RHD

Determinants	Effects	Impact on ARF and RHD burden
Socioeconomic & environmental factors 1. Poverty 2. Poor nutrition 3. Overcrowding 4. Poor standard of living	1. Rapid spread of GABHS 2. Difficulties accessing health care	1. Higher incidence of acute streptococcal pharyngitis and complications 2. Higher incidence of ARF and recurrent ARF
Health System Related Factors 1. Shortage of resources for health care 2. Low level of knowledge of disease among health care providers	1. Inadequate diagnosis and treatment of strep pharyngitis 2. Misdiagnosis or late diagnosis of ARF 3. Inadequate secondary prophylaxis delivery	1. Higher incidence of ARF and recurrent ARF 2. Missed first ARF episode 3. Inadequate secondary prophylaxis delivery 4. Higher rates of recurrent ARF with more frequent and severe

3. Low level of awareness of disease in the community		heart valve involvement 5. Higher rates of repeated hospital admissions and expensive heart valve surgery

2.3 Etiology and Pathogenesis

Rheumatic fever is thought to result from an inflammatory autoimmune response with antibodies produced against streptococcal antigen inducing inflammation in host tissue having similar molecules (ANTIGEN MIMICKERY THEORY). There is no direct invasion of tissues and toxin damage to tissues by the bacteria. Only group A beta-hemolytic streptococcal infections of the pharynx initiate or reactivate rheumatic fever.

In 0.3-3% of streptococcal pharyngeal infection, rheumatic fever develops several weeks after the sore throat has resolved. Studies show the existence of genetic predisposition in addition to bacterial factors.

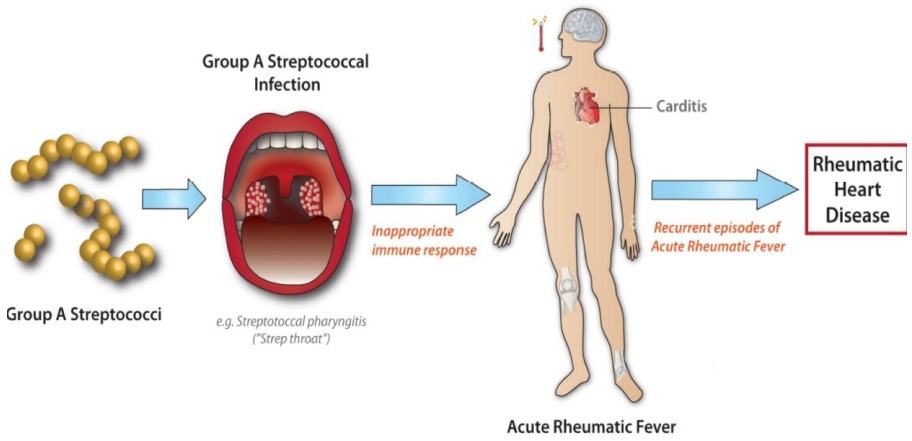


Fig 2.1. Pathogenetic pathway for ARF and RHD

Connective tissues, heart, brain and skin are the major organs involved by ARF. The symptoms and signs of ARF arise from involvement of these structures accompanied by systemic inflammatory reactions.

After recovery from the initial episode of RF, up to 60% to 65% of patients develop valvular heart disease and the risk of RF recurrence following GAS infection rises to 50%.

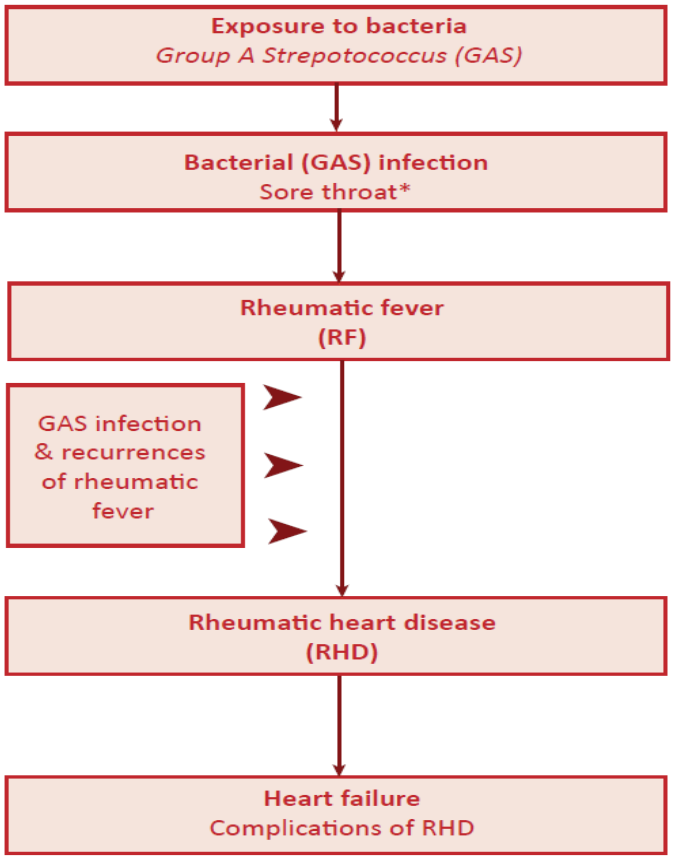


Figure 2.2 : Cascade of Sore throat, RF and RHD

Repeated GAS infections without appropriate treatment (with Benzathine Penicillin G) leads to RF recurrences and progressive valve damage-the defining characteristic of RHD which can, in turn, cause atrial fibrillation, heart failure, stroke and endocarditis.

2.4 Clinical Features

Following sore throat with GABHS, a silent period of 2 - 6 weeks precedes a sudden onset of fever, pallor, malaise and fatigue followed by characteristic major manifestations rheumatic fever start to appear which include the following:

- Arthritis
- Carditis manifests with tachycardia, tachypnea, dyspnea, chest pain, leg swelling,
- Sydenham's chorea with involuntary movement of extremities and the face
- Erythema marginatum
- Subcutaneous nodules

Other features that occur includes: Arthralgias Epistaxis, Serositis.

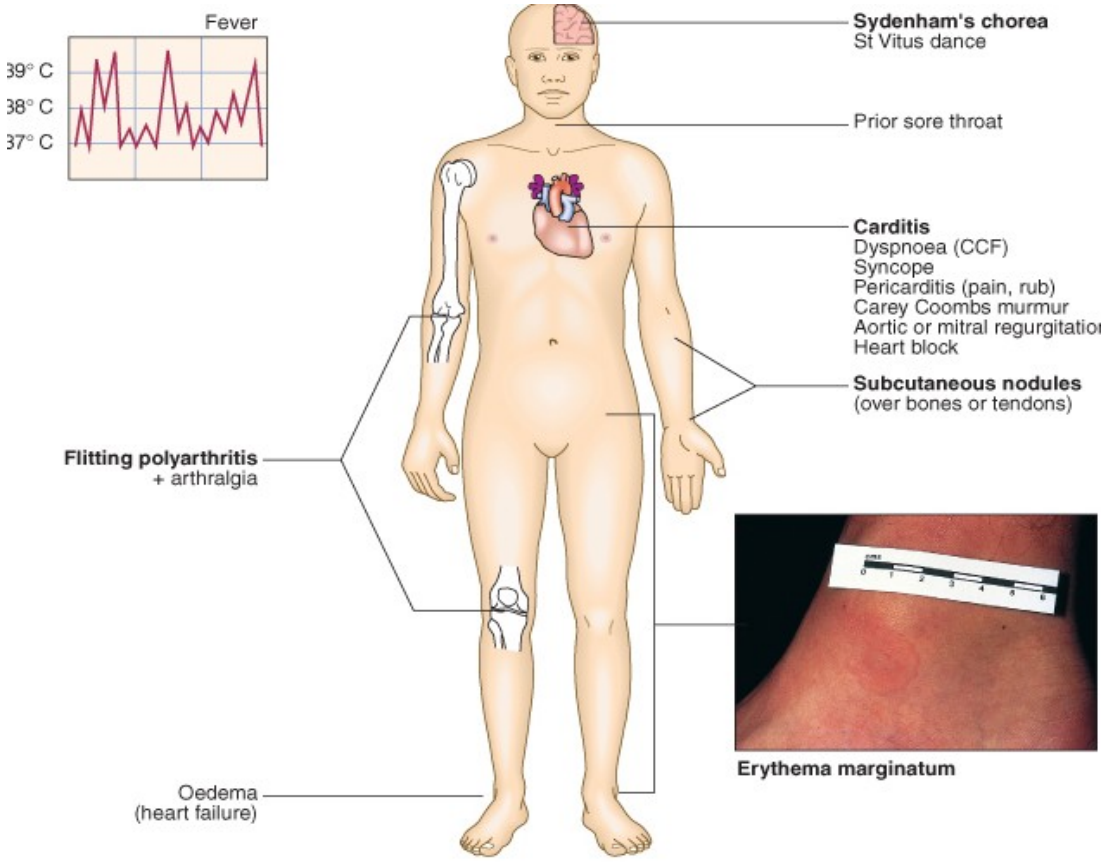


Figure 2.3: Classic manifestations of ARF

In one third of patients the streptococcal infection passes unnoticed and 54 to 70% of recurrences of ARF were caused by asymptomatic streptococcal infection.

2.4.1 Arthritis:

It's the earliest and most common symptomatic manifestation occurring in up to 75% of cases. It classically affects large joints like the knee, ankle, elbow and wrist in a migratory fashion (i.e. affecting one joint after the other) each being affected for 1-2 weeks period. Overall it lasts for up to six weeks and dramatically responds to anti-inflammatory agents like aspirin. Joints are hot, red, *tender*, swollen with limited mobility with asymmetric involvement. No residual deformity. It is unusual to involve the central joints as spines, hips and the peripheral ones as the fingers and toes. Infrequently it involves the temporo-mandibular joint.

It is more common in teenagers and adults than in children.

2.4.2 Carditis:

This is the most serious of all the manifestations of ARF which can lead to death in acute phase or at later stage. During the acute phase all the three layers of the heart are involved but later only the valves are affected. Carditis is the second common major manifestation occurring in 50-75% (40% during first attack and almost 100% in rheumatic fever recurrence) of ARF patients. (See RHD below).

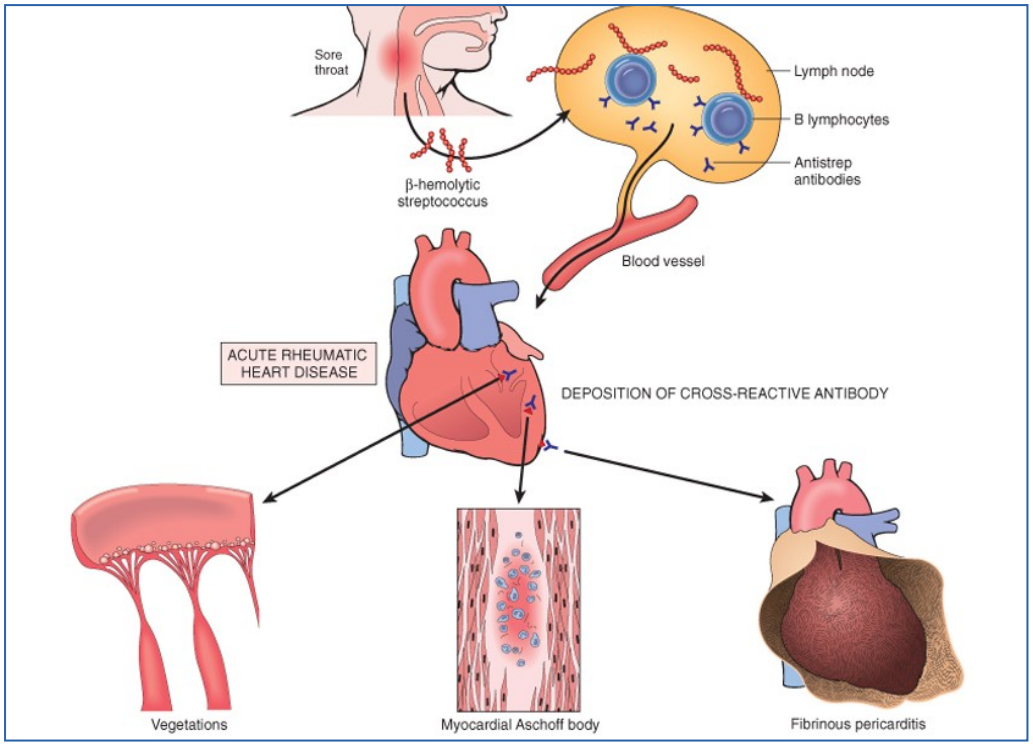
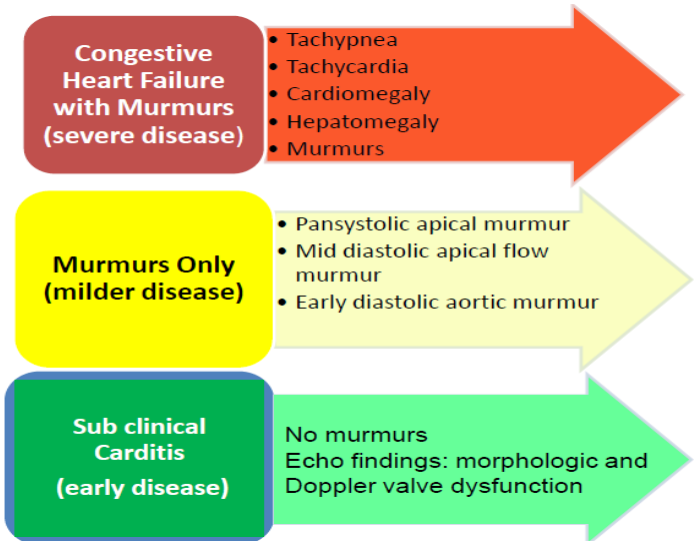


Figure 2.4: Pathogenesis of Carditis

It may be the only major manifestations and usually appears in the first week of the illness. Clinical features of carditis depend on severity of illness as follows:



2.4.3 Sydenham's Chorea :

It is a neurological disorder characterized by abrupt, semi-purposeful, non-rhythmic, repetitive dancing type of involuntary movements associated with muscular weakness and emotional changes. It subsides during sleep. Chorea has a longer latent period (up to 8 months), so other manifestations may be lacking or decreasing by the time Chorea develops for the first time. For this reason it can be diagnostic of rheumatic fever by itself. Chorea is due to basal ganglia involvement by ARF, affects

females more commonly and is self-limiting. It occurs in 10-20% of rheumatic fever patients.

Figure 2.5 Features of Chorea

2.4.4 Subcutaneous nodules:

This is mobile, painless nodule usually found on the extensor surfaces of joints of the hand specifically near or on the olecranon process. It's seen commonly in patients with carditis. It can occur with other diseases like rheumatoid arthritis.

They last for a week or two and rarely more than a month, and sometimes disappear within several days.

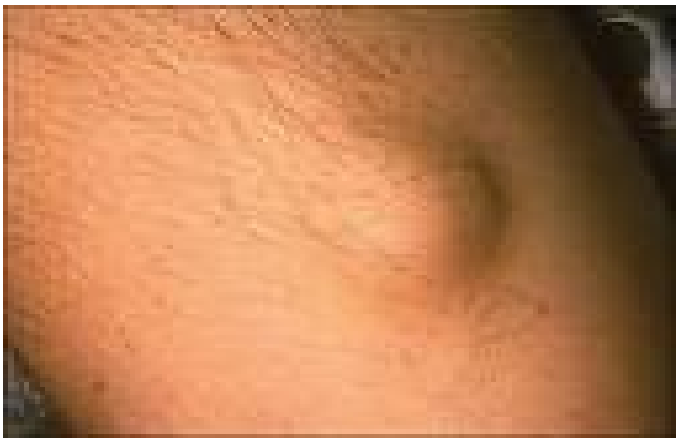


Figure 2.6: Subcutaneous nodules associated with rheumatic fever.

2.4.5 Erythema Marginatum

This is an erythematous, evanescent, non pruritic, non painful skin rash mainly found on the trunk and sometimes at the proximal part of the extremities like subcutaneous nodule. It usually occurs in the

covered parts and may be manifested by local application of heat. It is also seen commonly in association with acute carditis. They disappear within hours and may appear intermittently within weeks to months.

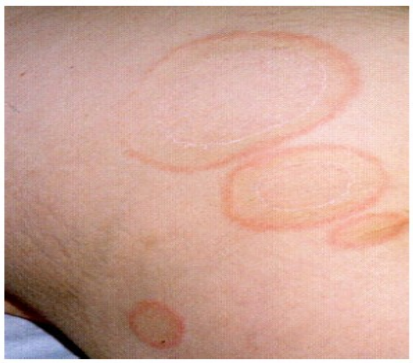


Figure 2.6: Erythema Marginatum

In summary the evolution of symptoms and signs are depicted in the figure below. The height indicates the frequency of the manifestation while horizontal dimension indicates the time of onset after the onset of systemic symptoms.

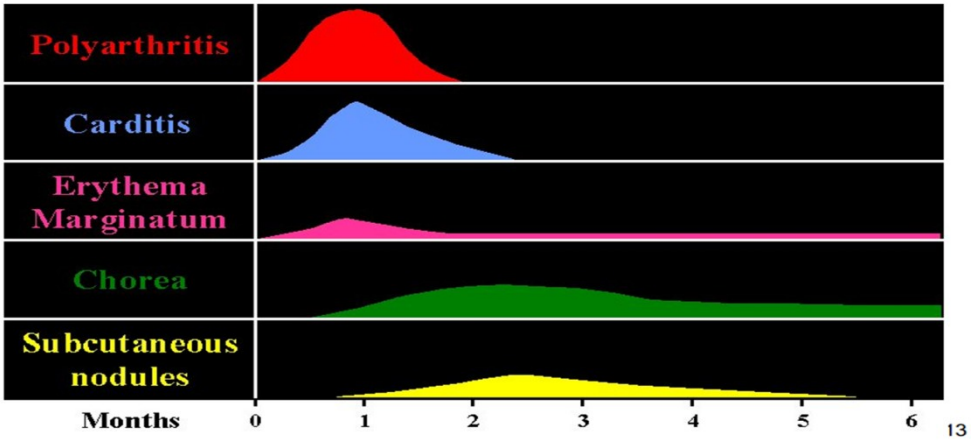


Figure 2.7 Clinical Manifestation of ARF in relation to time of onset

2.5 Laboratory findings in Acute Rheumatic Fever:

Elevated acute phase reactants

- Erythrocyte sedimentation rate(>30mm/hr)
- Leukocytosis
- C-reactive protein elevated

Recent Evidence of Group A Streptococcal infection:

- Raised ASO titer (80% of cases)
- Anti DNAase B
- Antihyaluronidase
- Rapid Stretococcal antigen test.

- Positive throat culture for (GAS),
- Recent scarlet fever

EKG: Increased **PR interval on EKG** (first degree heart block)

Chest X-ray-signs of congestion

Echocardiography-some valvular changes and regurgitations

2.6 Diagnosis:

There is no specific laboratory test to diagnose ARF and hence diagnosis is based on clinical criteria. The 2015 ACC/AHA ARF Revised Jones criteria for high prevalence countries (prevalence >1/1000 in all ages or incidence of 2/100,000 in school age children) are used to diagnose ARF in our country. For diagnosis of ARF the evidence of recent streptoccal infection should be demonstrated in addition to the criteria.

Table 2.2 Revised Jone's Criteria for Diagnosis of Acute Rheumatic Fever for high prevalence settings (2015 American College of Cardiology /American Heart Association).

<table>
<tr><td colspan="3">Evidence of preceding group A streptococcal infection (other than chorea):
✓ Raised ASO titer OR
✓ Positive throat culture for GABHS OR
✓ Positive Rapid antigen test OR
✓ Recent history of tonsilopharyngistis *strongly consider and emphasize</td></tr>
<tr><td rowspan="2">Diagnosis :</td><td>Initial ARF</td><td>2 major or 1 major plus 2 minor manifestations PLUS evidence of recent strep infection (other than chorea)</td></tr>
<tr><td>Recurrent ARF</td><td>2 major or 1 major and 2 minor or 3 minor PLUS evidence of recent strep infection (other than chorea)</td></tr>
<tr><td colspan="3">Criteria</td></tr>
<tr><td colspan="2">A. Major</td><td>B. Minor</td></tr>
<tr><td colspan="2">• Arthritis (Monoarthritis or polyarthritis or polyarthralgia[a]
• Carditis[b] (Clinical and/or subclinical)
• Chorea
• Erythema marginatum
• Subcutaneous nodules</td><td>• Monoarthralgia
• Fever (≥38°C)
• ESR ≥30 mm/h and/or CRP ≥3 mg/dL[c]
• Prolonged PR on ECG (for age) (unless carditis is a major criterion</td></tr>
</table>

[a]*Polyarthralgia should only be considered as a major manifestation after exclusion of other causes and in the presence of other major criterion. Joint manifestations can only be considered in either the major or minör categories but not both in the same patient.*

[b]*Sub-clinical carditis is pathological echocardiographic valvulitis*

[c]*CRP value must be greater than upper limit of normal for the laboratory. Also because ESR may evolve during the course of ARF, peak ESR values should be used.*

2.6.1 Diagnostic Classification of Acute Rheumatic Fever (ARF)

Definite ARF: 2 major, or 1 major plus 2 minor manifestations PLUS evidence of recent strep infection (other than chorea)

Highly Probable ARF: If an ARF diagnosis is considered highly probable (but not confirmed due to lack of evidence for recent streptococcal infection). This group may be more common in Ethiopia as there is often lack of laboratory tests to confirm recent streptococcal infections.

Uncertain ARF: in patients from high-risk groups with only one major manifestation of acute Rheumatic fever or borderline echocardiographic findings.

2.7 MANAGEMENT OF ACUTE RHEUMATIC FEVER

Persons with symptoms of ARF should be referred to a hospital. The purpose of the referral is to ensure accurate diagnosis, and to give clinical care and education about preventing further episodes of ARF.

Observation and confirmation of ARF is required prior to anti-inflammatory treatment, however paracetamol may be given for fever or joint pain.

Certain investigations should be done to rule out other potential causes for the symptoms and signs and to build the case for ARF.
Investigations includes CBC, ESR, CXR, EKG and ASO Titer (if available)
An initial echocardiogram (if available) is important to help identify and measure heart valve damage.
Once diagnosis is confirmed, long-term preventative management is organized before discharge.

Once the diagnosis of ARF is decided management has several components:

A. Treat the Infection:

All newly diagnosed cases of ARF should receive antibiotics

- A single intramuscular injection of benzathine penicillin G (BPG) to eradicate GAS from upper respiratory tract.
    - ✓ 600 000 IU for those < 30kg and
    - ✓ 1.2 million IU for those > 30kgs.
- Erythromycin PO for 10 days, if allergic to penicillins.

After this initial course of antibiotic therapy the patient should be started on long term monthly BPG secondary prophylaxis.

B. Treat Arthritis and fever

Use paracetamol at doses of 60 mg/kg/day in four divided doses until the diagnosis is confirmed. Mild arthralgia and fever may respond to paracetamol alone. Arthritis and severe arthralgia require treatment with nonsteriodal anti-inflammatory drugs (NSAIDs) like Aspirin, ibuprofen or Naproxen.

Dosing:

- ✓ Start **Aspirin** 75 mg per kilogram per day divided 6 hourly after meals for up to 4-6 weeks , OR
- ✓ **Ibuprofen** 30mg/kg per day 8 hourly.

After initiation do ESR 2 weekly and taper aspirin dose by decreasing the dose by 2 tablets every week.

The duration of treatment is dictated by the clinical response and improvement in inflammatory markers (ESR, CRP). Many patients need anti-inflammatory therapy for only 1–2 weeks (i.e. anti-inflammatory therapy can be stopped at 2 weeks if the patient is pain free with improved inflammatory markers). In some patients, joint symptoms may recur following the cessation of treatment (so-called 'rebound phenomenon'); this does not indicate recurrence, and can be treated with another course of anti-inflammatory therapy.

If Patient is not responding or not tolerating aspirin:

- ✓ Start Prednisolone 2mg per kilogram per day for 2 weeks; then aspirin is added at dose 60 mg per kilogram per day divided into 4 doses for another 2 weeks; then Prednisolone is tapered & discontinued.
- ✓ Do ESR 2 weekly, taper aspirin by decreasing the dose by 2 tablets every week.

Most ARF episodes subside within 6 weeks, and 90% resolve within 12 weeks. Approximately 5% of patients require 6 months or more of anti-inflammatory therapy.

C. Treat Carditis

- ✓ Bed rest if in heart failure, with mobilization as symptoms permit
- ✓ Do Urgent echocardiogram
- ✓ Manage heart failure:
  - Diuretics/fluid restriction for mild or moderate failure
  - Furosemide 1-2mg/kg PO/day
  - ACE inhibitors like enalapril or Lisinopril for more severe failure, particularly if AR is present
  - Prednisolone may be given for severe carditis

- Anti-coagulation medication and digoxin if atrial fibrillation is present
- Valve surgery for life-threatening acute carditis (rare)

D. Treat Chorea

- ✓ Most mild-moderate cases do not need medication
- ✓ Provide calm and supportive environment (prevent accidental self-harm)
- ✓ Carbamazepine at 7–10 mg/kg day in three divided doses or Valproic acid at 15–20 mg/kg/day in three divided doses or phenobarbitone at 3-6 mg/kg/day (particularly in children) can be given for severe cases.

E. Other management considerations

- ✓ The patient's vital signs should be recorded four times daily, and the pattern and extent of fever noted and follow laboratory parameters. The patient should be examined daily for the pattern of arthritis, and the presence of heart murmur, choreiform movements, skin rash and subcutaneous nodules.
- ✓ Register patient in a RHD Register
- ✓ Ask about family members: those with sore throat are given one injection of benzathine penicillin or oral antibiotics for 10 days.
- ✓ Educate client and family on Sore throat, ARF, RHD, Family planning, dental care, importance of secondary prophylaxis and regular medical follow up.

F. Management of probable Rheumatic Fever

Patients with probable ARF may be managed in two ways according to the level of confidence with which the diagnosis is made:

i. ***Highly-suspected ARF:*** *manage as for definite ARF*
If an ARF diagnosis is considered highly probable (but not confirmed due to lack of evidence for recent streptococcal infection) management should be commenced as for definite ARF .

ii. ***Uncertain ARF:*** in patients from high-risk groups with only one major manifestation of acute Rheumatic fever or borderline echocardiographic findings , administer 12 months of secondary prophylaxis initially, and reassess (including echocardiography) at that time. If there is no evidence of recurrent ARF, and no evidence of cardiac valvular damage on echocardiography at 12 months, consider ceasing secondary prophylaxis.

G. Advice on discharge from Hospital

- All patients should have a good understanding of the cause of rheumatic fever and the need to have sore throats treated early.

- Family members should be informed that they are at increased risk of ARF compared to the wider community.
- Patients and families should understand the reason for secondary prophylaxis and the consequences of missing a BPG injection.
- They should be given clear information about where to go for secondary prophylaxis, and written information on appointments for follow up with their local health professional, medical practitioner, Internist/Pediatrician and cardiologist (if needed).
- They should be given contact details for the RHD register coordinator (if there is one), and encouraged to telephone if they have any questions concerning their follow up or secondary prophylaxis.
- They should also be reminded of the importance of antibiotic prophylaxis for dental and other procedures to protect against endocarditis.

2.7 Prevention of ARF:

Depends on eradication of group A streptococci from upper respiratory tract.

1. Primordial Prevention:

Improving socioeconomic conditions, nutrition, housing conditions (decreasing crowding) and improving access to health care can all decrease the incidence of ARF.

2. Primary prevention:

In patients with a sore throat and symptoms suggestive of a GAS infection, antibiotic treatment using intramuscular BPG could reduce the risk of RF by up to 80% .

However, about 1/3 of patients with ARF do not recall preceding episode of pharyngitis

A vaccine for GAS is being developed but has not yet been used in clinical practice

3. Secondary Prophylaxis:

Monthly injection of BPG IM to prevent recurrences of rheumatic fever

Case study 1

A 16 year old boy presented to the health facility with a sore throat 1 month back. His throat was very red and painful during swallowing. The health professional gave him paracetamol for pain relief and sent him home.

He returned to clinic a day back with a painful right hip and swollen right hand, headache, and fever (38.2 ^{o}C). The health professional thinks he can hear a strange heart beat sound. His ESR is 42mm/hr. His WBC count is 6000/mcl. He gives no personal history or family history of ARF or RHD.

Q1. What is the likely diagnosis and what is the immediate treatment?

Time allocation

- Group discussion - up to 15 mins
- Presentation and feedback to each group – 10 mins per group (max 30 mins)
- General discussion by the facilitator - up to 5 mins.

Case study 2

A 13 year old girl presents to a health facility with strange movements in her hands and face. She cannot poke her tongue out straight; and she cannot touch her nose with her finger. You also notice that she has a strange gripping movement of her right hand when she tries to hold something. She doesn't give any reported history of any illness and is otherwise healthy. All routine blood tests including CBC and ESR are normal.

Q1. What is the likely diagnosis and

Q2. What should be the immediate treatment?

Time allocation

- Group discussion - up to 15 mins
- Presentation and feedback to each group – 10 mins per group (max 30 mins)
- General discussion by the facilitator - up to 5 mins

Unit 2 Summary

- ARF is an autoimmune consequence of infection with Group A streptococcal infection.
- It results in a generalized inflammatory response affecting brains, joints, skin, subcutaneous tissues and the heart.
- The clinical presentation can be vague and difficult to diagnose.
- Currently the Revised Jones criteria ACC/AHA 2015 forms the basis of the diagnosis of the condition.
- Acute rheumatic fever is managed by addressing and controlling the acute inflammatory process through Non-steroidal anti-inflammatory particularly ASA .
- A long-term Management Plan should be established to prevent recurrence of ARF and development or worsening of RHD Probable ARF cases should also be monitored.

UNIT 3: SECONDARY PROPHYLAXIS IN RHEUMATIC FEVER AND RHD

Learning Objectives

At the end this module the participants will be able to:-

- Discuss secondary prophylaxis of ARF
- Explain Safe BPG injection practices
- Manage penicillin allergy and anaphylaxis
- Discuss secondary prophylaxis options

3.1 Introduction

Secondary prevention refers to the early detection of disease and implementation of measures to prevent recurrence and worsening disease. Term used to describe regular delivery of antibiotics to prevent recurrence of GAS infection and subsequent development of ARF.

3.2 When should secondary prophylaxis be considered?

- ARF confirmed by the Revised Jones Criteria
- RHD confirmed on echocardiogram
- ARF or RHD not confirmed but considered highly 'probable'
- RHD post surgery

NB: Continue prophylaxis in pregnant patients and those on warfarin.

3.3 Benefits of Secondary prevention in RF/RHD

Secondary Prevention:

- Prevent further Group A Streptococcal infections
- Prevent recurrence ARF
- Prevent the development of RHD
- Reduce the severity or worsening of RHD
    - It is associated with regression of heart disease in approximately 50-70% of those with good adherence over a decade and reduces mortality.

3.4 Standard Drugs, Dose and Frequency

A. 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 4 weeks. Benzathine penicillin G is should be given by deep intramuscular injection every 4 weeks.

The standard dose is:

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

B. Erythromycin

Erythromycin is given if there is a proven allergy to Penicillin. The standard dose is **250mg oral, twice-daily for children < 7 yrs and 500mg PO BID for ALL people > 7 years of age.**

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

3.5 Penicillin allergy and anaphylaxis

Allergic reactions to Benzathine penicillin G injections are rare and fatal reactions are even 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.

Studies by the International Rheumatic Fever Study Group **(IRFSG)** in 1988-90 and National RF/RHD prevention and control programme from 32 hospitals of Nepal from 2007-2010 have shown that the prevalence of penicillin allergy and anaphylaxis is very low.

Table 3.1 Prevalence of Penicillin Reactions

Reactions	**International Rheumatic Fever Study Group 1991**	**Nepalese Heart Journal Vol.8 | No. 1 | October, 2011**
Allergic reaction	57/1790 people (**3.2%**)	60/4712 patients **(1.3%)** (of 77300 injections)
Anaphylaxis	4/1790 people (**0.22%**)	5/4712 patients (**0.1%**)
Death	1/1790 people (**0.05%**) Patient with Severe Mitral Valve Disease and Heart failure.	No death was reported
Vasovagal reaction		8/4712 patients **(0.16%)** (6 of them were with severe RHD).

Benzathine penicillin G (and Penicillin V) should not be given to persons with a proven serious (anaphylactic) penicillin allergy. 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 fetus 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 bleeding following injections despite a normal INR.

Patients and health care providers have several concerns on use of Benzathine Penicillin G.

Table 3.2: Potential barriers to BPG Use and suggested solutions

Problem	Solution
The drug is ‘’Heavy”	Use Appropriate amount of diluents at room temp.
The drug can block the needle	Use a large bore needle
The injection is painful	Dilute the powder in lidocaine and inject slowly
Patients fear allergy	Serious allergy is very rare-reassure patients
Health workers fear allergy	Training of health workers on allergy management help them to be confident
Not understanding benefits of BPG	Educate on relation between sore throat and RHD

3.6 How to administer Benzathine Penicillin

Benzathine Penicillin G use recommendations:

1. Benzathine Penicillin G (BPG) is an essential medicine for treatment and for prevention of ARF/RHD
2. Alternatives to BPG (e.g. oral medicines) are not as effective as BPG
3. Serious allergic reactions to BPG are rare and should not stop us from using BPG.
4. BPG injection can be given by any trained health worker following standard procedures as shown below.
5. Skin testing with dilute BPG will not predict the patients who are allergic, therefore it is not indicated.
6. Inform patient or guardian and obtain informed verbal consent.
7. Follow the 5 steps protocol below.

The following figure shows the 5 steps protocol that needs to be followed for administration of Benzathine Penicillin G.

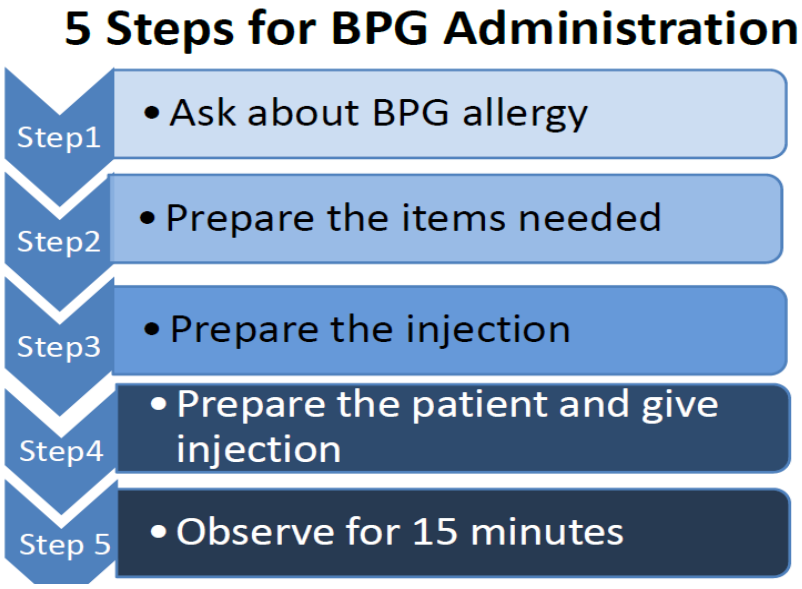
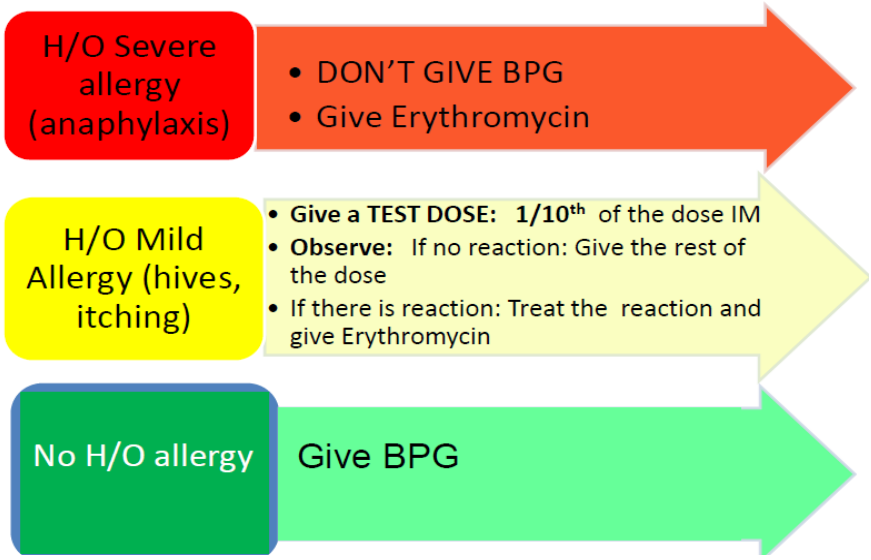


Figure 3.1: the 5 Steps for BPG Administration
(Adapted from Sudan RHD Guidelines 2017)

STEP 1: Ask about the History of Allergy and decide



STEP 2: Prepare the items needed

1. One 10 ml syringe
2. One 5 ml syringe
3. One BPG ampoule 1.2 million units
4. One vial of local anesthetic lidocaine 2% (or water for injection)
5. One adrenaline vial 1:1000
6. One antihistamine vial: Promethazine 50mg inj

7. Normal saline 1000ml: 1 bag (with IV Cannula)

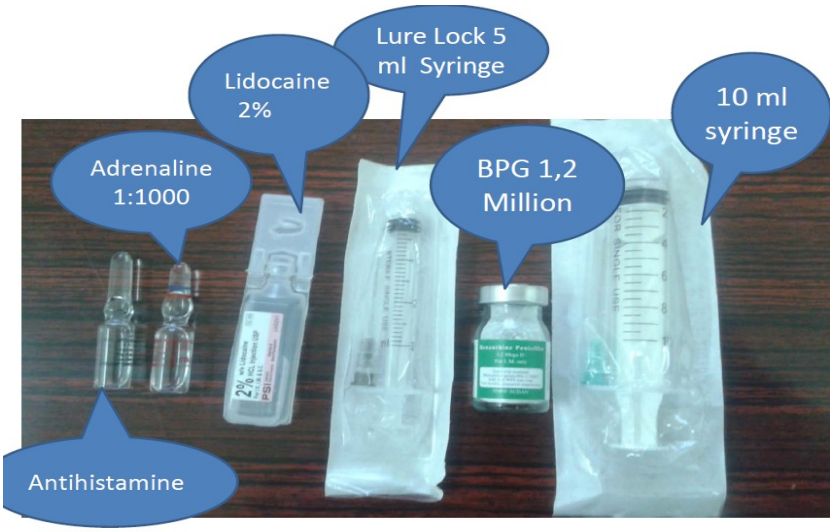


Figure 3:1 Items needed for BPG Injection

STEP 3: Prepare the injection:

1. Draw appropriate amount of local anesthetic as diluents for the BPG powder or water for injection if no lidocaine(make sure it's not cold)
2. Inject the diluents into the BPG vial
3. Mix gently till dissolved by rolling in the hands
4. Draw in 5 ml syringe
5. Change the needle to a large bore (10 ml syringe) needle.

STEP 4: Prepare the Patient and Give the Injection:

1. Ask the patient to lie on the abdomen
2. Mark the site of the injection on the gluteus muscle
3. To minimize pain: press with your thumb over the site for 10 seconds
4. Aspirate first to avoid veins then give the injection SLOWLY deep in the muscle.
5. Use new needle for each patient
6. Discard used needles and syringes in safety box
7. Keep the patient for 15 minutes
8. Document the date and dose on the patient chart and the patient passport

- **NEVER EVER GIVE BPG INTRAVENOUSLY AS THIS MAY LEAD TO IMMEDIATE DEATH!!**
- **SKIN TESTING IS NOT RECOMMENDED TO CHECK FOR ALLERGY TO BPG.**

STEP 5: Observe and treat reactions

Observe the patient for at least 15 minutes.

If reaction any develops evaluate the patient and classify the reaction as follows and act accordingly:

A. Mild Reaction (Local Reaction) :
   - Itching, hives or urticaria : manage with antihistamine injection. Continue observation until the patient is well

B. Severe Reaction (Anaphylaxis):
   - sudden face/tongue swelling with difficulty breathing, BP < 90/60 or collapses over minutes to hours

C. Vasovagal (Pain) Reaction:
   - sudden immediate collapse and transient loss of consciousness usually in a very sick patient.

3.7 Management of Penicillin reactions

3.7.1 Vasovagal Reaction

Vasovagal reaction is a transient loss of consciousness due to a reflex response that encompasses vasodilatation and/or bradycardia, leading to systemic hypotension and cerebral hypoperfusion. This reaction may occur due to pain of penicillin injection or emotional stress. It occurs more in patients with severe RHD.

Prevention of Vasovagal Reactions:

Pain is the main factor contributing to Vasovagal reactions hence injection techniques that help reduce pain will decrease Vasovagal reactions.

- Use a smaller-gauge needle: a 21-gauge needle
- Warm syringe to room temperature immediately before using.
- Ensure that skin swabbed with alcohol is dry before injecting
- Apply gentle pressure to the injection site for 10 seconds with the finger or thumb before injection
- Deliver injection slowly (preferably over at least 2–3 min)
- Distract patient during injection (e.g. with conversation)
- The addition of 0.5–1 mL of 1% lignocaine may help
- Keep the patient supine during the injection

Management of Vasovagal reaction:

- Protect patient from falls and injuries.
- Patients should be advised to assume the supine position with legs raised at the onset of symptoms.
- Advise the patient to do Isometric Counter-pressure maneuvers :
  - Leg-crossing with simultaneous tensing of leg, abdominal, and buttock muscles

- Handgrip, which consists of maximum grip on a rubber ball or similar object
- Arm tensing, which involves gripping one hand with the other while simultaneously abducting both arms

- Educate and reassure the patient and family.
- Closely monitor vital signs

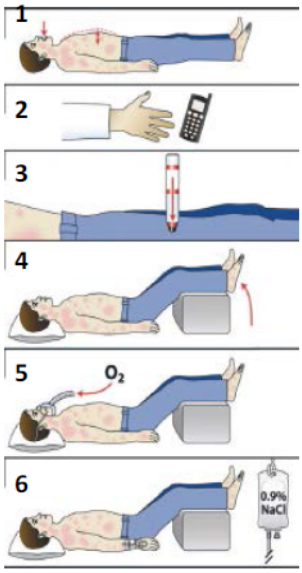
3.7.2 Anaphylaxis

Diagnosis:

Anaphylaxis is highly likely when any one of the following 3 criteria are fulfilled:

1. Acute onset of an illness (minutes to several hours) with involvement of the skin, mucosal tissue, or both (eg, generalized hives, pruritus or flushing, swollen lips-tongue-uvula)
   AND AT LEAST ONE OF THE FOLLOWING
   a. Respiratory compromise (eg, dyspnea, wheeze-bronchospasm, stridor, reduced PEF, hypoxemia)
   b. Reduced BP or associated symptoms of end-organ dysfunction (eg, hypotonia [collapse], syncope, incontinence)
2. Two or more of the following that occur rapidly after exposure to a likely allergen for that patient (minutes to several hours):
   a. Involvement of the skin-mucosal tissue (eg, generalized hives, itch-flush, swollen lips-tongue-uvula)
   b. Respiratory compromise (eg, dyspnea, wheeze-bronchospasm, stridor, reduced PEF, hypoxemia)
   c. Reduced BP or associated symptoms (eg, hypotonia [collapse], syncope, incontinence)
   d. Persistent gastrointestinal symptoms (eg, crampy abdominal pain, vomiting)
3. Reduced BP after exposure to known allergen for that patient (minutes to several hours):
   a. Infants and children: low systolic BP (age specific) or greater than 30% decrease in systolic BP*
   b. Adults: systolic BP of less than 90 mm Hg or greater than 30% decrease from that person's baseline

Management of Anaphylaxis:



1.Assess ABC: if needed perform CPR

DO STEPS 2,3,4 QUICKLY AT SAME TIME

2.Call for help

3.Inject adrenaline: 0.3 ml (<7 y), 0.5 ml (>7 y) of 1:1000 solution. Can be repeated in 15 minutes

4.Lie the patient with legs up

5.If distressed, give O2

6.IV line: give IV normal saline10ml /kg boluses. Can give adrenaline infusion

World Allergy Organization Guidelines for anaphylaxis management. WAO2011;4:13-37

3.8 Duration of Secondary Prophylaxis

Different guidelines recommend different lengths of secondary prophylaxis. Since Ethiopia is a high prevalence country with suboptimal RHD Care the Ethiopian Society of Cardiac professionals recommend the following prophylaxis regimen.

Table 3.3: Duration of recommended secondary prophylaxis

Disease Classification	**Duration of Secondary Prophylaxis**
ARF without carditis	1. Minimum of 5 years after last ARF, or 2. Until age 21 years (*whichever is longer)*
ARF with carditis but no current residual carditis	1. Minimum 10 years after last ARF, or 2. Until age 25 years (*whichever is longer)*
RHD and following **Cardiac Surgery** for RHD	Continue medication for life

3.9 Documentation and adherence support

Recording and reporting of secondary prophylaxis is a key component of RHD Prevention and control program.

Record in the Benzathine penicillin injection book and/or medical notes

- Dose and batch number
- Date given and date next due
- Signature (*of person giving injection*)
- Record next date due on a reminder card (if applicable)

Educate the patient about the importance of adherence to monthly BPG injections

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

However,

- If injections were PRESCRIBED for the full year but none were GIVEN, record 0%, OR
- Receiving less than 50% of injections places an individual at extreme risk of recurrent ARF and progression of RHD.

Immediate intervention is required for these individuals.

Strategies to improve BPG Injection Delivery

- Appoint a dedicated staff member at each clinic to oversee secondary prophylaxis coordination
- Identify people who need secondary prophylaxis
- Identify local health facility for each person
- Develop systems for follow-up
- Provide ongoing education for people who require injections and their families
- Communicate with local RHD programme and other health service providers

- Reduce injection pain
- Discuss alternative therapy issues

UNIT 3 Summary

- Antibiotics need to be present in the body at all times to help prevent GAS infections and prevent recurrent ARF
- Benzathine penicillin injections should be given unless there are contraindications to injections or documented severe penicillin allergy.
- Medical Specialist review is required before ceasing secondary prophylaxis
- Strategies to improve secondary prophylaxis delivery:
  - Good relationships between community and health staff
  - Education for the community and health staff
  - Systems for follow-up
  - Communication between health services
  - Reduce injection pain
- Document Benzathine Penicillin injections and monitor injection delivery

Unit 4: Diagnosis and Management of Rheumatic Heart disease

Learning Objectives

At the end this module the participants will be able to:-

- Describe burden of RHD
- Describe pathogenesis of RHD
- Discuss the clinical features of RHD
- Describe the methods of diagnosis of RHD
- Discuss management of RHD

4.1 Introduction

RHD is inflammation of heart valves that follows infection with Group A beta hemolytic streptococcus, commonly pharyngitis.

Rheumatic disease (RHD) is the only long term sequelae of ARF which can lead to disability or death It is thought that 40-60% of patients with ARF will go on to developing RHD. Rheumatic carditis affects mainly the heart valves. Histopathology may show the Aschoff nodules (the characteristic lesions of RHD) in the myocardium and pericardium however clinical symptoms and signs are mainly due to valvular involvement.

4.2 Pathophysiology of RHD

Involvement of the valves is the fundamental pathophysiologic change of RHD which affects mainly the mitral and aortic valves. History of Rheumatic fever can be elicited in less than half of patients who come for the first time with RHD.

In most patients RHD remains clinically silent (10- 20 years) during which the valve lesion progresses to cause significant valvular abnormalities due to recurrent attacks of unrecognized Acute Rheumatic Fever.

Heart valves normally allow easy forward blood flow from one chamber of the heart to another by adequate opening and prevent back flow by appropriate complete closure.

Progression of the initial inflammation results in fibrotic changes of the valves interfering with normal function of the valves.

In patients with RHD permanent damage of the valve apparatus leads to scarring resulting in poor opening and closure of the valves.

If valves fail to properly open to allow adequate forward flow narrowing (**stenosis**) of the valves is said to have occurred.

If the valves fail to properly close backward flow or leakage of blood (**regurgitation**) will develop.

Patients can have isolated stenosis or regurgitation or combined stenosis and regurgitation.

The mitral valve is the most common valve involved followed by aortic, tricuspid and very rarely pulmonary valve in that order.

4.3 Clinical Manifestations of RHD

4.3.1 Symptoms

Symptoms of RHD may not develop for many years

- ✓ A murmur but no symptoms usually suggests mild-moderate disease
- ✓ Symptoms usually suggest moderate-severe disease

Symptoms depend upon the type and severity of disease, and may include

- ✓ Breathlessness on exertion (dyspnea) or when lying down flat(orthopnea)
- ✓ Waking at night feeling breathless (paroxysmal nocturnal dsypnoea)
- ✓ Feeling tired
- ✓ Generalized weakness
- ✓ Palpitations
- ✓ Peripheral edema
- ✓ left anterior chest pain and syncope in addition to heart failure symptoms can also occur

4.3.2 Physical Examination

Clinical assessment should be conducted carefully because early detection of RHD can result in a better outcome. Patients with RHD may have the following findings if they come with heart failure or other complications

- ✓ Tachcyardia(Increased heart rate)
- ✓ Tachycapnia(increased respiratory rate)
- ✓ Raised temperature
- ✓ Cyanosed lips and tongues (Bluish discoloration)
- ✓ Chest Crackles
- ✓ Heart Murmurs
- ✓ Tender RUQ area (because of liver congestion)
- ✓ Leg edema
- ✓ Weak extremities because of stroke.
- ✓ Chest crackles and cardiac murmurs

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 (See table below).

Table 4.1 Common Murmurs in RHD

Valve affected	Description
Mitral regurgitation	A pansystolic murmur heard loudest at the apex and radiating laterally to the axilla
Mitral stenosis	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.
Aortic regurgitation	A diastolic blowing decrescendo murmur best heard at the left sternal border with the person sitting up and leaning forward in full expiration.
Aortic stenosis	A loud, low pitched mid-systolic ejection murmur best heard in the aortic area, radiating to the neck.

4.4 Diagnosis of RHD

Acute Rheumatic carditis with other manifestation of Acute Rheumatic Fever is diagnosed by Jones Criteria 2015 mentioned above. Unfortunately most patients come to health institution at late stage of Rheumatic heart disease (Chronic Rheumatic heart Disease) with heart failure or complications like stroke or infective endocarditis . It is not also uncommon for some women to present for the first time during pregnancy . It is only few patients who give history of ARF when they come with heart failure or complications.

Accurate diagnosis of RHD requires echocardiographic detection of valve abnormality and hemodynamic changes like regurgitation and ejection fraction. Unfortunately echocardiography is not available widely and clinical suspicion is therefore very important to early diagnose and prevent the progression of RHD.

In general all patients having heart failure symptoms or manifestations which look like complications of RHD should be referred to the next level of health care for better diagnosis and/or echocardiographic assessment by more qualified health professionals (cardiologist, Internist or pediatrician).First step is to make sure that there is no rheumatic fever recurrence or infective endocarditis.

Laboratory and other Investigations in RHD

- ✓ CBC and ESR
- ✓ Electrocardiogram (ECG)
    - To check for rate and rhythm
- ✓ Chest X-ray (CXR)
    - To determine size and placement of heart
    - To look for signs of heart failure (pulmonary congestion)

- ✓ Echocardiography
    - To identify heart valve damage
    - To estimate severity of disease
    - Useful to compare results with future echocardiogram results.

4.5 Management of RHD

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.

The key principles for effective management of RHD include:

- Effective baseline assessment, education and referral
    - Establishing the diagnosis of RHD (see above)
    - Detecting and treating Complications of RHD (see below)
- Treatment of cardiac and other symptoms
- Long-term secondary prophylaxis (to prevent recurrent ARF)
- Regular medical and cardiology review including echocardiography
- Appropriate and timely surgical interventions
- Dental assessment and care
- Advise on Family planning and referral
- Management of RHD in special situations*(e.g . pregnancy)*

4.5.1 Medical Management of RHD

It encompasses the identification and management of complications of RHD and management of patient after cardiac interventions:

- Heart failure
- Atrial fibrillation
- Infective endocarditis
- Stroke
- Post cardiac intervention care

4.5.2 Valve surgery or interventions for RHD

The need for surgery and cardiac interventions in RHD depends on:

- Severity of symptoms
- Evidence that the heart valves are severely damaged
- Left ventricular chamber size and ejection fraction
- Availability of long-term management after surgery *(i.e. anticoagulation)*

Indications for interventions

1. Asymptomatic Patients:

Asymptomatic patients with severe valvular lesion (see Echo criteria for severity) should be closely monitored to decide the appropriate time for intervention.

The following are indications for intervention:

- Severe Pulmonary hypertension.
- For regurgitant lesions (MR and AR), decrease in LV ejection fraction or Increasing in LV dimensions especially LV end systolic dimension.
- Severe MS.

2. Symptomatic patients:

- Symptomatic patients with severe valve dysfunction should be referred for intervention.

Type of Interventions in Rheumatic Heart disease

The Heart valves can be repaired or replaced. Choice of intervention should take into account availability of facilities, future pregnancy, age and ability to continue to take anticoagulation.

For Patients who wishes to be pregnant and unable to take oral anticoagulant, valvotomy, valve repair or tissue type of prosthetic valve should be considered.

For those with atrial fibrillation, younger age, able to take oral anticoagulation and pregnancy is not an issue mechanical valve replacement should be considered.

Cardiac Intervention options

1- Valve repair

- This is done mainly for MR but needs careful assessment by Echocardiography to select the suitable candidates.

2- Percutaneous transvalvular mitral commissurotomy (PTMC):

- Patients with severe MS and suitable valve anatomy should be considered for PTMC or for closed mitral valvotomy (CMV), if PTMC is not available.

3- Valve replacement:

- Prosthetic valve replacement is the option when valve repair or commissurotomy is not feasible for MV disease and it is the main procedure for AV disease.
- Valve replacement can be either mechanical valve or bioprosthetic valve.

Assessment before surgery includes

- Echocardiogram to assess severity of heart valve damage
- Complete dental assessment and treatment *(if required)*

- Review and management of other health problems *(e.g. Kidney disease, vascular and chronic respiratory disease, cancers, malnutrition or obesity)*

Post Valve Surgery Care

Valve surgery is not curative for RHD and many complications can happen after valve surgery so patients need continuous follow up care. It includes:

- Continue Benzathine Penicillin G monthly injections for life
- In patients with mechanical prosthetic valves:
  - Warfarin should not be stopped
  - INR control: target 2.5-3
- Endocarditis prophylaxis before high risk procedures
- Dental hygiene
- Regular medical and echocardiographic review.

Table 4.2: Recommended routine review and management plan for ARF and RHD

Classification	Criteria	Review and Management Plan	Frequency
High Risk (Priority level 1)	Severe valvular disease **or** Moderate/severe valvular lesion with symptoms **or** Mechanical prosthetic valves, tissue prosthetic valves and valve repairs, including balloon valvuloplasty	Secondary prophylaxis	3-4 weekly
		Doctor/HO/BSC Nurse review	3-monthly
		ECG (optional)	Yearly
		Medical or Heart specialist review	6-monthly
		Echocardiogram	Annually for surgery or post surgery valve status assessment
		Dental review	Within 3 months of diagnosis , yearly thereafter
		Endocarditis prevention	As required
		Warfarin or Aspirin	As prescribed
Medium Risk (Priority level 2)	Any moderate valve lesion in the absence of symptoms, and with normal left ventricular function	Secondary prophylaxis	4 weekly
		Doctor/HO/BSC Nurse review	6-monthly
		ECG (optional)	Yearly
		Medical or Heart specialist Review	Yearly
		Echocardiogram	Every 1 years for children Every 2 years for adults
		Dental review	Yearly
		Endocarditis prevention	As required
Low Risk (Priority level 3)	ARF with no evidence of RHD ***or*** Trivial to mild	Secondary prophylaxis	4 weekly
		Doctor/HO/BSC Nurse review	Yearly
		Dental review	Yearly
		Medical or Heart	Every 2 years for adults.

	valvular disease	specialist Review	Yearly for children<18 years of age
		Echocardiogram (if available)	Every 2 years for children Every 3 years for adults

Unit 4 Summary

- After attack of ARF some patients progress to develop progressive valvular damage due to repeated attacks of ARF which could be noticed or unnoticed.
- Progressive valvular damage by RHD can interfere with proper closure or opening of the heart valves resulting in narrowing (stenosis) or can interfere with proper closure resulting in Regurgitation or both.
- The symptoms of RHD depend on the valve lesion and its severity.
- Clinical assessment should be conducted carefully because early detection of RHD can result in a better outcome.
- Accurate diagnosis of RHD requires echocardiographic detection of valve abnormality.
- The mitral valve is most commonly affected, followed by Aortic, Pulmonary and rarely Tricuspid valves
- RHD may be asymptomatic or it can have mild, moderate or severe symptoms
- Management of RHD includes
    - Treatment of heart failure and other symptoms
    - Long-term secondary prophylaxis (to prevent recurrent ARF)
    - Regular medical and cardiology review
    - Management of existing pregnancy
    - Dental assessment, family planning referral .

Unit 5: Management of Complications of Rheumatic Heart Disease

Learning objectives

At the end this module the participants will be able to:-

- Describe common Complications of RHD
- Identify and refer patients with complications
- Explain the management of common complications of RHD
- Discuss the approaches in management of RHD in special conditions

5.1 Introduction

As discussed in earlier sections of this training RHD causes damage to the valves causing scarring and leading to either narrowing of the valve orfice which will impede forward flow of blood and/or may make the valve not able to close properly resulting in leakage of blood backwards. These morphologic or functional changes on the valve when it progresses will result in heart failure.

Damaged valves are also at increased risk of infection resulting in what is called infective endocarditis.

Patients with RHD are also at increased heart rhythm abnormality (arrhythmia). The most common arrhythmia seen in patients with RHD is atrial fibrillation.

Infective endocarditis and arrhythmias can precipitate heart failure in patients with RHD and also predispose to development of blood clots in the heart (thrombus) which can embolize to different parts of the body (eg. to the brain and cause stroke, or to peripheral arteries and cause gangrene in extremities).

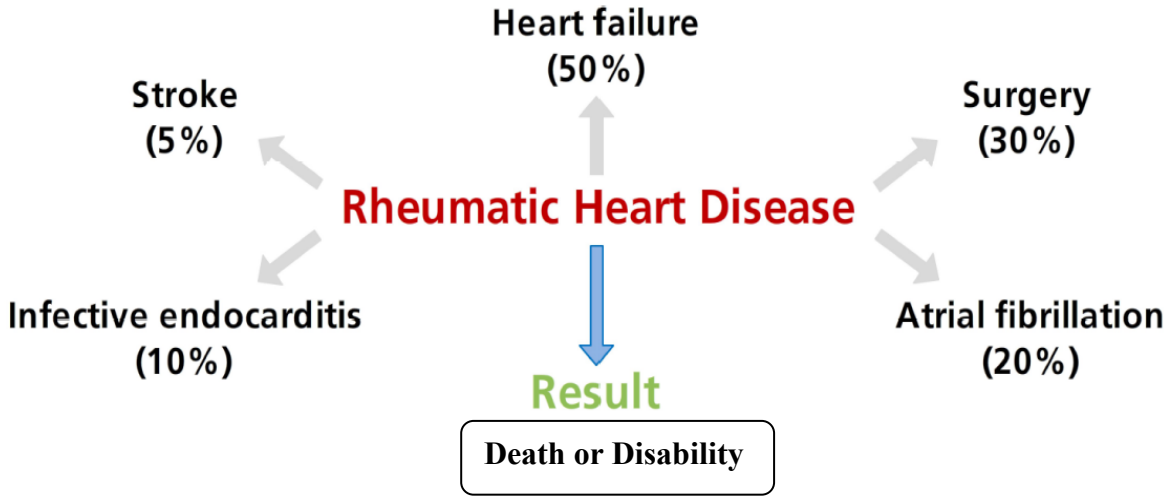


Figure 5.1 Long term sequelae of RHD

5.2 Heart Failure

5.2.1 Introduction

Heart failure is constellation of symptoms and signs arising from structural or functional cardiac disorders that impair the ability of the ventricle(s) to fill with and/or eject blood.

Abnormal filling will result in accumulation of fluid (congestion) in tissues like in lung, abdomen and extremities causing shortness of breath and edema.

Abnormality in pumping adequate blood will result in poor perfusion of tissues and cause fatigue (exercise intolerance), syncope and angina.

RHD can result in either filling abnormality or poor pumping of the heart by causing abnormal valvular function.

Heart Failure is the most common presentation of RHD in Ethiopia. The initial manifestation of heart failure usually comes with some precipitating causes which put extra load on the heart. The most common precipitating causes of heart failure in RHD include:

* Fever
* Anemia
* Infective endocarditis
* Rheumatic recurrence
* Atrial Fibrillation
* Excess salt intake
* Pregnancy

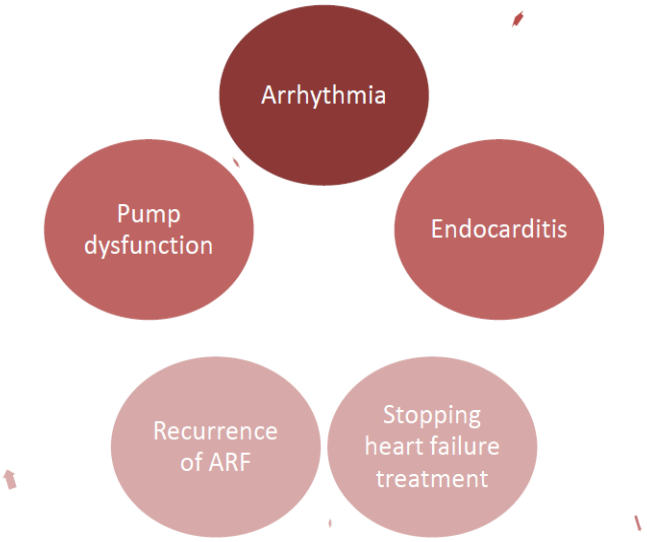


Figure 5.2 Causes of Decompensation/Worsening Heart Failure in RHD

5.2.2 Pathophysiology of Heart Failure in RHD

Clinically, heart failure presents with features of pulmonary congestion or left heart failure (LHF) initially, then it may progress with time to develop pulmonary hypertension (PHT) and right sided heart failure (RHF) with features of both LHF and RHF i.e. congestive heart failure (CHF) then with

the progression of Pulmonary hypertension the features of pulmonary congestion may diminish and the picture becomes that of severe RHF and end stage cardiac disease.

Fluid retention in HF is initiated by the fall in cardiac output, leading to alterations in renal function, due in part to activation of the sodium-retaining renin-angiotensin-aldosterone and sympathetic nervous systems.

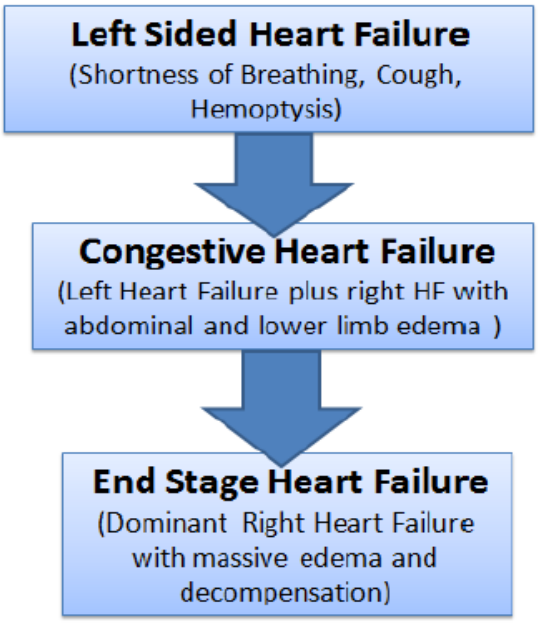


Figure 5.3 Pathophysiology of HF in RHD

5.2.3 Clinical Manifestations of Heart Failure

The clinical manifestations of heart failure arise from either excess fluid accumulation or reduced cardiac output (See table 5.1)

Table 5.1: Clinical Manifestations of heart failure

Symptoms due to excess fluid accumulation	Symptoms due to a reduction in cardiac output	Physical Findings in heart failure
• cough, • wheezing, • dyspnea, • orthopnea, • paroxysmal nocturnal dyspnea, • lower extremity edema, • abdominal pain from hepatic congestion, • Ascites(abdominal distension)	• fatigue, • weakness, • decreased exercise tolerance • palpitations • fainting/syncope	• Tachcyardia (Increased heart rate) • Third heart sound besides the “lub” and “dub”(S3 gallop) • Irregular or alternating pulse • Tachypnea (increased breathing rate) • laterally displaced apical impulse; • Heart murmurs • Elevated jugular venous pressure or positive hepato-jugular reflux • Cyanosed lips and tongues (bluish discoloration)

		• Tender RUQ area (because of liver congestion) • Peripheral edema • Chest crepitations

5.2.4 Workup of Suspected heart failure

Refer patients with suspected heart failure to hospital for further work up and initiation of treatment. The initial evaluation generally includes

- chest radiography- check for increased cardiothoracic ratio, congestion,
- electrocardiography- ECG is always abnormal in heart failure,
- echocardiography- to check for valves, measure ejection fraction, regurgitation or pulmonary hypertension ,
- laboratory tests: complete blood count, ESR, Cr, SGOT, SGPT, serum potassium and Sodium.

5.2.5 Diagnosis and Grading of Heart Failure

The diagnosis of heart failure is a complex process but for primary health care level the modified Framingham criteria is used as a diagnostic tool (table 5.2).

Table 5.2: Modified Framingham clinical criteria for the diagnosis of heart failure

(Source: UpToDate 2017)

Major Criteria	Minor Criteria
• Paroxysmal nocturnal dyspnea	• Bilateral leg edema
• Orthopnea	• Nocturnal cough
• Elevated jugular venous pressure	• Dyspnea on ordinary exertion
• Pulmonary rales	• Hepatomegaly
• Third heart sound	• Pleural effusion
• Cardiomegaly on chest x-ray	• Tachycardia (heart rate ≥120 beats/min)
• Pulmonary edema on chest x-ray	• Weight loss ≥4.5 kg in five days
• Weight loss ≥4.5 kg in five days in response to treatment of presumed heart failure*	
Diagnosis The diagnosis of heart failure requires that **2 major or 1 major and 2 minor criteria** that cannot be attributed to another medical condition.	

Grading Severity of Heart Failure: As heart failure is a dynamic state that needs different management, grading of the severity is very important. We use New York Heart Association and Modified Ross Heart Failure Classification for Children for this purpose.

Table 5.3: Heart Failure Classification-New York Heart Association for Adults and Modified Ross for Children

Class	NYHA(for Adults)	Modified Ross (for Children)
Class I	No limitations of physical activity	No limitations or symptoms
Class II	May experience fatigue, palpitations, dyspnea, or angina during moderate exercise but not during rest	**Infants**: Mild tachypnea or diaphoresis with feeding **Older children**: Mild to moderate dyspnea on exertion
Class III	Symptoms with minimal exertion that interfere with normal daily activity	**Infants**: Growth failure and marked tachypnea or diaphoresis with feeding **Older children**: Marked dyspnea on exertion
Class IV	Unable to carry out any physical activity because they typically have symptoms of HF at rest that worsen with any exertion	Symptoms at rest such as tachypnea, retractions, grunting, or diaphoresis

5.2.6 Management of Heart Failure

Initial management of heart failure depends on severity of symptoms and concomitant conditions

- Mild Symptoms (class II and III):
  - Bed rest and salt restriction
  - Oral Diuretics titrated to symptoms.
- Patients with severe decompensation should be managed at hospital level where specific diagnosis can be made and close monitoring is possible.
  - NYHA/ROSS Class IV heart failure
  - Pulmonary edema
  - Disabling right side congestive heart failure.
  - Cardiogenic Shock

Patients with heart failure should be referred for identification of underlying causes and initiation of heart failure medications. Stabilized patients can be followed at health center .

The basic components of heart failure management includes:

A. Manage the Congestive state

The management of the congestive state requires addressing three hemodynamic parameters that affect the cardiac output (See table 5.4). These are;

- Reduce Preload

- Increase contractility
- Reduce Afterload

Table 5.4: Treatment to decrease the congestive state in heart failure

Decrease Preload	Increase contractility of the heart	Decrease Afterload:
• Decrease salt intake to less than 2g/day (don't add salt to foods) • Give Diuretics: o Furosemide with a initial dose of 1-2mg/kg PO escalated to a dose which can achieve a weight loss of 0.5-1kg/day for edematous/congested patient and then tapered down to lowest dose which can make the patient symptom free. Start at 40mg PO/day in adults. Escalate based on Wt., urine output ,response and lab tests (to max 400mg/day in adults).	• Indicated for those with reduced contractility (Ejection fraction less than 60% in patients with Mitral regurgitation and less than 40% in all other patients. • Digoxin is the usual drug given to enhance contractility. Dose 0.125-0.25mg PO/day.	• Indicated for patients with severe acute MR or AR and patients with Chronic severe MR or AR for whom surgery is not possible. • Commonly used drug is Enalapril 2.5 mg PO BID to max of 10mg PO BID

B. Identify and treat precipitating causes

It is important to identify and treat any reversible factors, which may be exacerbating the symptoms of HF. Patients being evaluated for exacerbation of HF should be thoroughly assessed for any factor(s) that may have contributed to the exacerbation. These factors include:

- ❖ Anemia
- ❖ Arrhythmia: Atrial fibrillation
- ❖ Infective endocarditis
- ❖ Infections such as pneumonia
- ❖ Rheumatic recurrence
- ❖ Excessive salt intake
- ❖ Poor adherence to drugs
- ❖ concomitant drugs such as NSAIDs
- ❖ Pregnancy

C. Retard progression of the heart damage

- Benzathine penicillin is given to avoid the progression of the underlying heart disease by decreasing recurrence of rheumatic fever.
- In some patients where the valve lesion has advanced and looks like features of dilated cardiomyopathy add ACE inhibitors, spirinolactone (dose: 12.5 to 25 mg PO/Day) and beta blockers can be given to avoid excessive remodeling.
- Prevention of Infective endocarditis

D. Treat the underlying cause in RHD.

These are not easily and timely available currently in Ethiopia.

- ❖ Surgery: Valve replacement or repair
- ❖ Valvotomy: usually done for Mitral stenosis

Follow up of Heart failure management

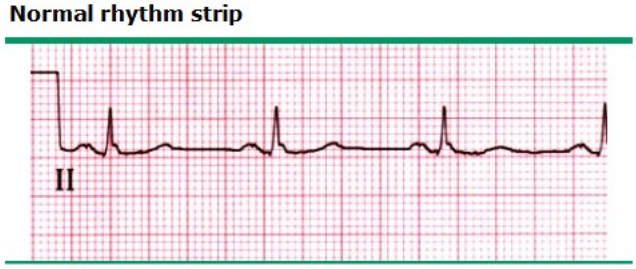
In the management of heart failure monitor the following parameters:

- Worsening of symptom (fatigue, shortness of breath and body swelling).Symptoms should be graded with NYHA functional class. Any deterioration in functional status requires referral.
- Occurrence of complications like infective endocarditis, stroke or cardioemolism.
- Occurrence of edema,
- heart rate ,rhythm, blood pressure
- Weight
- Monitoring urine out put, serum electrolytes and creatinine in admitted patients

5.3 Atrial Fibrillation

Patients with RHD are at increased risk of developing heart rhythm abnormalities. The most common arrhythmia is atrial fibrillation. An irregularly irregular pulse is suggestive of atrial fibrillation. Atrial fibrillation is commonly seen in patients with mitral stenosis.

Patients having irregular heart rhythm should be referred to hospitals to confirm the diagnosis and initiate treatment for AF and anticaogualtion



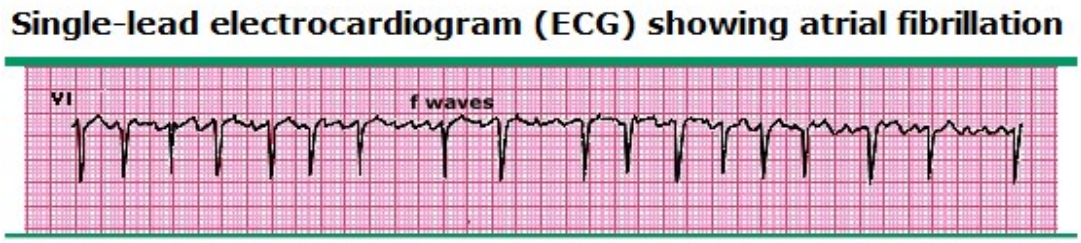


Figure 5.4: Normal ECG and ECG showing Atrial fibrillation

The most feared complication of atrial fibrillation is the development of cardiac thromboembolism (formation in the heart and dissemination of thrombus through the systemic circulation). The thrombus can embolize to the:

- Brain causing stroke (sudden weakness of extremities)
- Extremities causing peripheral arterial disease (sudden onset of limb pain ,darkish discoloration of the limb(gangrene)

Atrial fibrillation causes abrupt loss of the atrial contribution to ventricular filling and as much as a **30 percent reduction in cardiac output** and generally a rapid ventricular rate with associated shortened diastolic filling time. This may precipitate or aggravate heart failure.

Management of Atrial fibrillation in RHD

Management should address the atrial fibrillation and its potential complications.

There are two options for management of atrial fibrillation: rate control versus rhythm control.

We prefer rate control for RHD as there is no clear survival benefit of rhythm control over rate control and it is costly and needs higher expertise.

i. Rate control:

If patient does not have pulmonary congestion or hypotension beta blockers PO like metoprolol 25-100mg/day or atenolol 50-100mg/day can be given to keep heart rate between 60-80/min at rest and 90-115 beats/min with exercise If however, the patient has pulmonary congestion or hypotension digoxin 0.125-0.25mg/day PO can be given to control the heart rate.

Patients with heart failure and coexisting atrial fibrillation should be treated with diuretics and ACE inhibitors and this will often reduce the heart rate.

ii. Anticoagualtion in RHD and AF

Anticoagulation is indicated in patients with RHD and:

- atrial fibrillation or
- history of embolization (stroke) and
- following valve replacement (Bioprosthetic or Mechanical)

Medicines and Monitoring:

- Vitamin K antagonists like warfarin are the preferred drugs.
- Usual starting dose is Warfarin 2.5mg /d and then escalated based on INR. It usually takes 3-5 days to know the effects of the new dose.
- **INR Target**: 2.0 -3.0
- **Frequency of INR** -Once baseline INR is determined, initially patients should be monitored weekly until they achieve target INR and then monthly once target INR is achieved.
- Patients should be advised on symptoms of excessive anticoagulation like epistaxis, easy bruising, delayed stopping of bleeding after needle puncture or minor trauma, reddish discoloration of urine, heavy menses or easy bruising.

NB: even though Aspirin is not the most effective medicine in the prevention of cardioembolism,in situations where warfarin is not available or is not safe, Aspirin 160-325mg PO/day can be used.

5.4 Infective Endocarditis

Infective endocarditis (IE) is an important complication of heart disease in general and valvular heart disease in particular.

As RHD constitutes the major cause of valve heart disease in Ethiopia, it is the most commonly encountered heart disease predisposing to IE.RHD was found to be the underlying cardiac lesion in 49% of patients seen in a cohort of children with IE, and the mortality in this cohort was 7.3%. Congestive heart failure and systemic embolization occurred in 66% and12% respectively.

This is infection of the damaged valves by microbial agents which have got access to the blood stream In RHD, endocarditis most commonly occurs in the mitral or aortic valves. The most common organisms causing IE arise from normal flora like from gingival tissue . The most common focus of such infection is the periodontal region . If organisms get access to the blood stream they can easily cause infection of the valves resulting in infective endocarditis. Uncommonly occurs during dental or surgical procedures but often the source of the infection is not clear. It may occur after heart valve surgery.

Diagnostic Clues:

Fever, hematuria, night sweating,weight loss, clubbing, changing murmur, unexplained rapid deterioration of heart failure, splenomegaly. Skin manifestations like conjunctival hemorrhage, splinter hemorrhage. Infective endocarditis is far less common in pure stenotic lesions. Though history of predisposing procedures are commonly enquired it is not commonly elicited in most of our patients.

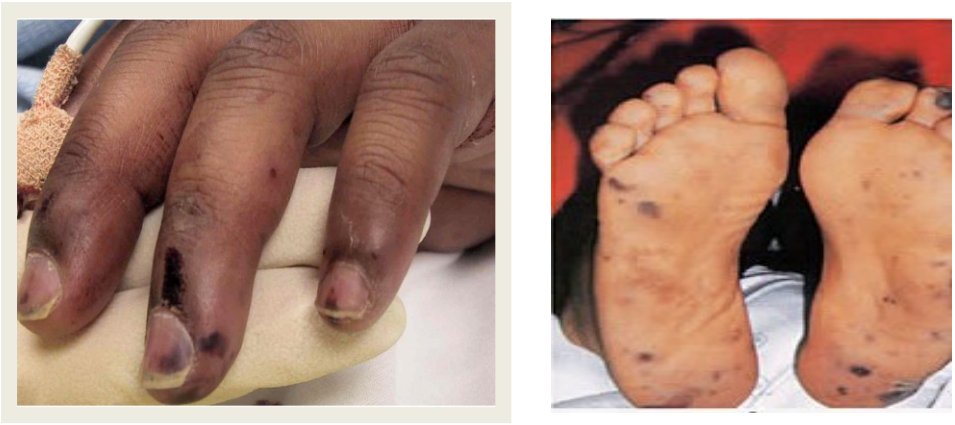


Figure 5.5 Skin manifestations in IE A: Splinter hemorrhages B. Janeway Lesions

Laboratory Features in Infective Endocarditis:

1. Anemia
2. Most commonly elevated WBC
3. ESR elevated,
4. Microscopic hematuria
5. Bacteremia

Diagnosis of Infective Endocarditis:

Diagnosis should be guided by the Modified Duke Criteria (Table 5.5)

Table 5.5: Modified Duke Criteria for Diagnosis of Infective endocarditis

Major criteria:

1. **Positive Echocardiogram**:

   - Oscillating intracardiac mass on valve or supporting structures in the path of regurgitant jets or on implanted material in the absence of an alternative anatomic explanation.
   - Intramural abscess.
   - New partial dehiscence of a prosthetic valve.

2. **Histopathological** evidence of IE from excised heart valve.
3. Positive **blood culture** with an organism consistent with IE.
   - Typical micro-organisms in 2 separate cultures or
   - Persistently +ve blood cultures drawn 12 hours apart or
   - Single +ve blood culture of Coxiella burnetti

Minor criteria

1. Predisposing heart condition (e.g. RHD) or IV drug use.
2. Fever.

3. **Vascular phenomena**: major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages and Janeway lesions.
4. **Immunologic phenomena**: gluomerulonephritis, Osler nodes, Roth's spots and positive rheumatoid factor.
5. **Microbiological evidence**: positive blood culture, but does not meet a major criterion.
6. **Echocardiographic** abnormalities that fell short of typical lesions described above.

Decision Criteria:

Definite IE:2 Major or 3 Minor + 1 Major or 5 Minor

Possible IE: 1 Major +1 Minor, Or 3 minor

Rejected IE: firm alternative Diagnosis or response to <4 days of antibiotics.

In contrast to what is stated in literature, blood culture for unknown reason is often negative. So treatment is basically empiric based on strong clinical suspicion and echocardiographic findings of vegetation.

Empiric Treatment of Infective Endocarditis:

All patients suspected of having or confirmed to have infective endocarditis should be referred to hospital for admission and evaluation by cardiologist or internist/pediatrician.

Take adequate blood samples for culture before initiation of antibiotics.

Choice of Antibiotics:

After taking blood culture, the patient should be started on IV antibiotics:

- Crystalline penicillin 3millin Units IV 4 hourly, OR Ampicillin 2 g IV 4 hourly for 4-6 weeks.
- plus gentamicin 1mg/kg IV tid for 2 weeks.

Ceftriaxone 2g IV/day can be used in place of penicillins but it doesn't cover enterococci.

In case of prosthetic valve endocarditis, the drugs should include vancomycin (15mg/kg BID IV)

In patients presenting with toxic manifestations and of short duration or patients who continued to have fever after initiation of empiric antibiotics for five days staphylococcus aureus and other resistant organisms should be considered and Vancomycin with aminoglycoside or Rifampcin should be started.

Surgical intervention may be needed if there is no improvement by medical treatment or there is a high risk of embolization (size, site and mobility of vegetation)

Patients should be monitored clinically and with some laboratory parameters like ESR, urinalysis and hemoglobin and if available blood culture

Dental Care and Infective Endocarditis Prophylaxis in RHD

This requires emphasis on improved access to dental care and improved oral health in patients with underlying cardiac risk factors for infective endocarditis, rather than a sole focus on dental procedures and antibacterial prophylaxis.

Optimal oral health is maintained through regular professional care and the use of appropriate products such as manual and powered toothbrushes, floss and other plaque-control devices such as antibacterial mouthwashes.

Patients need to be strongly advised to comply with a continuing oral and dental care regimen and should be referred annually for dental check up.

Treatments to achieve this goal include:

- Removal of impacted teeth and unerupted teeth
- Treatment of all teeth with periapical disease by endodontic debridement and root filling or apical surgery or extraction.
- Removal of all carious teeth that cannot be restored.
- Treatment of other abnormalities such as cysts or intra-bony lesions associated with the dentition and related structures
- Treatment of oral ulcers including those caused by ill-fitting or irritating dental appliances
- Treatment of inflammatory periodontal disease like gingivitis.
- Oral hygiene instructions for the patient to ensure maintenance of ideal oral health.

Dental procedures (plus tonsillectomy/adenoidectomy) for which endocarditis prophylaxis is recommended

- All dental procedures that involve manipulation of gingival tissue or the periapical region of teeth or perforation of the oral mucosa
- Endocarditis prophylaxis is no longer recommended for non-dental procedures (including respiratory, gastrointestinal and genitourinary procedures)

Cardiac conditions for which endocarditis prophylaxis is recommended

- Prosthetic heart valves (bioprosthetic or mechanical)
- Rheumatic valvular heart disease
- Previous endocarditis
- Unrepaired cyanotic congenital heart disease (includes palliative shunts and conduits)
- Surgical or catheter repair of congenital heart disease within 6 months of repair procedure.

Antibacterial regimen for dental procedures (plus tonsillectomy/adenoidectomy)

Amoxicillin 2g (child: 50mg/kg up to 2g), administered

- Orally, 1 hour before the procedure, or
- IV, just before the procedure, or
- IM, 30 minutes before the procedure.

For **penicillin allergy or if a penicillin or cephalosporin-group antibiotic is taken** more than once in the previous month (including those on long-term penicillin prophylaxis for rheumatic fever):

Clindamycin 600mg (child: 15mg/kg up to 600mg), administered

- Orally, 1 hour before the procedure, or
- IV, over at least 20 minutes, just before the procedure, or
- IM, 30 minutes before the procedure. Or

Clarithromycin 500mg (child: 15mg/kg up to 500mg) administered orally, 1 hour before the procedure.

5.5 Special considerations: RHD and Pregnancy

Normal pregnancy is associated with a 30–50% increase in blood volume, reduction in systemic vascular resistance and corresponding increase in cardiac output. These changes begin during the first trimester, peaking at 28–30 weeks of pregnancy, and are then sustained until term. The increase in blood volume is associated with an increase in heart rate by 10–15 beats per min. Because of the hyperdynamic circulation, innocent, soft mid-systolic murmurs are common during pregnancy, particularly along the left sternal border.

These changes can exacerbate any pre-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. Sometimes RHD, especially mitral stenosis, is first diagnosed during pregnancy or soon after delivery when a woman develops symptoms, usually dyspnea.

Ideally, women with known rheumatic valve disease should be fully assessed before pregnancy occurs so that any necessary intervention may be safely done. Women at particularly high risk may be counseled to avoid pregnancy like in severe pulmonary hypertension, severe aortic stenosis or patient who had NYHA class IV heart failure.

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 in pregnancy generally includes:

- restricting physical activity and decreasing salt intake;
- administering appropriate secondary prophylaxis*(Benzathine penicillin should be continued during pregnancy)*;
- avoiding community-acquired infectious diseases or institute early treatment;
- education about monitoring own signs and symptoms and seeking care if shortness of breath occurs;
- 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

- Severe mitral and/or aortic stenosis;
- Atrial fibrillation;
- Prosthetic heart valves;
- Those receiving anticoagulant therapy with warfarin
- Severe Pulmonary Hypertension.

Discuss benefits and risks of anticoagulation during pregnancy and manage anticoagulation.

Risks of anticoagulation in Pregnancy and post partum in RHD

Maternal:

1. Hypercoagulable state of pregnancy can lead to bleeding
2. Thromboembolism

Fetal:

1. Miscarriage
2. Fetal embryopathy

Management of anticoagulation

- Avoid warfarin in first trimester (teratogenic).
- Use Heparin in first trimester and after 37 weeks
- Stop anticoagulants during labor.
- Anticoagulation is reinstituted following delivery.

Case Study 1

Alemitu is a15 year old student brought to health centre by her parents for she complained fatigue, shortness of breath on going to toilet . She also gives history of cough with intermittent blood mixed sputum. Alemitu's teacher told her parents that Alemitu couldn't do physical education as her peers in her class and she got easily tired since six months back. On examination she was found to have respiratory rate of 32/min, heart rate of 124/min, BP of 100/70, distended neck veins, murmur on auscultation of the left anterior chest , Right upper quadrant tenderness and bilateral leg edema.

Questions:

1. What additional history would you like to get from Alemitu?
2. What is the likely diagnosis?
3. What is her functional class?
4. What investigations are important to confirm the diagnosis?
5. Discuss the management of the case.
6. What advice would you give to Alemitu's parents

Case Study 2:

Mihretu is 25 year old teacher suddenly failed to use his right hand while writing on black board and also unable to move his right leg. He then started to talk with signs but was able to understand what people talk. He was immediately taken to nearby health center and the health worker found the following findings: BP of 130/85, PR of 120/min (irregularly irregular), clear chest , loud murmur at apex of the heart and weak right upper and lower extremity with deviation of the mouth to the left.

Questions:

1. What additional history would like to obtain?
2. What is the likely diagnosis of the case?
3. Discuss the mechanism of development of the problem in relation to the primary problem.
4. What other sites of the body can be involved with such presentations?
5. What investigations are required to confirm the diagnosis and manage the case?
6. How do you manage the patient?

Unit 5 Summary

- Heart failure, atrial fibrillation, stroke and infective endocarditis are common complications of RHD.
- Heat failure is a clinical syndrome arising from inability of the heart to **pump** adequate cardiac output or inability to **fill** with normal filling pressure
- Heart failure is treated with diuretics to decrease fluid accumulation in tissues and drugs to boost contraction of the heart like digoxin and drugs like ACE inhibitors to retard progression of the heart muscle damage.
- Management of Atrial fibrillation is essential to prevent heart failure as well as embolic stroke using beta blockers, digoxin and warfarin.
- Embolic complication of rheumatic heart disease are prevented through anticoagulation.
- The management of Infective endocarditis involves high index of suspicion for early diagnosis and use of parenteral antibiotics for 4 to 6 weeks.
- Pregnancy complicated by RHD may be detrimental to mother or the baby hence appropriate referral and management should be instituted.

Unit 6: Rheumatic Heart Disease Prevention and Control Program

Learning Objectives

At the end this module the participants will be able to:-

- Explain why RHD program is important for Ethiopia.
- Clarify the World Heart Federation comprehensive RF/RHD Prevention and control framework
- List the important components of an RHD Prevention and control Program

6.1 Introduction

The following are some of the reasons why we need a RHD prevention and control program.

- Morbidity and mortality due to RHD is very high.
- RHD is a preventable condition acquired in childhood that is amenable to early and effective intervention.
- Prevention is cheaper and cost effective
- RHD care is also an issue of equity as it disproportionately affects the poor.
- Advanced care including surgery is expensive
- Routine care and attention given to RHD is very low.
- Awareness of the community and the health care workers on sore throat, rheumatic fever and RHD is low

Based on the World Heart Federation Position Statement the goal of RHD prevention is To reduce premature deaths from rheumatic fever (RF) and RHD among individuals aged <25 years by 25% by the year 2025.

6.2 Rheumatic fever and RHD Prevention and Control Framework

The World Heart Federation Roadmap for Reducing CV Morbidity and Mortality Through Prevention and Control of Rheumatic Heart Disease published a continuum of care model for RHD (Figure 6.1). According to that model people with streptococcal tonsillo-pharyngitis ('strep throat'), RF and RHD share a number of common health care needs.

An unwell individual or their caregiver must initially decide to seek care for the illness (CoC2), and then engage with the health system (CoC3) where they should be appropriately diagnosed (CoC4) either with strep throat (path 1), rheumatic fever (path 2) or rheumatic heart disease (path 3). Those with strep throat should receive appropriate treatment with intramuscular benzathine penicillin G (BPG), ideally at the primary care level (CoC6). Those diagnosed with rheumatic fever should begin secondary prophylaxis using BPG (CoC7) also at the primary care level, and be referred for appropriate investigation at higher levels. And those with RHD should be referred (CoC5) for

appropriate treatment at higher levels of care (CoC8), after which they should receive the appropriate follow-up care (CoC9 and CoC10). The framework also underscores the roles that poverty and the social determinants of health play as crucial risk factors for RHD (CoC1).

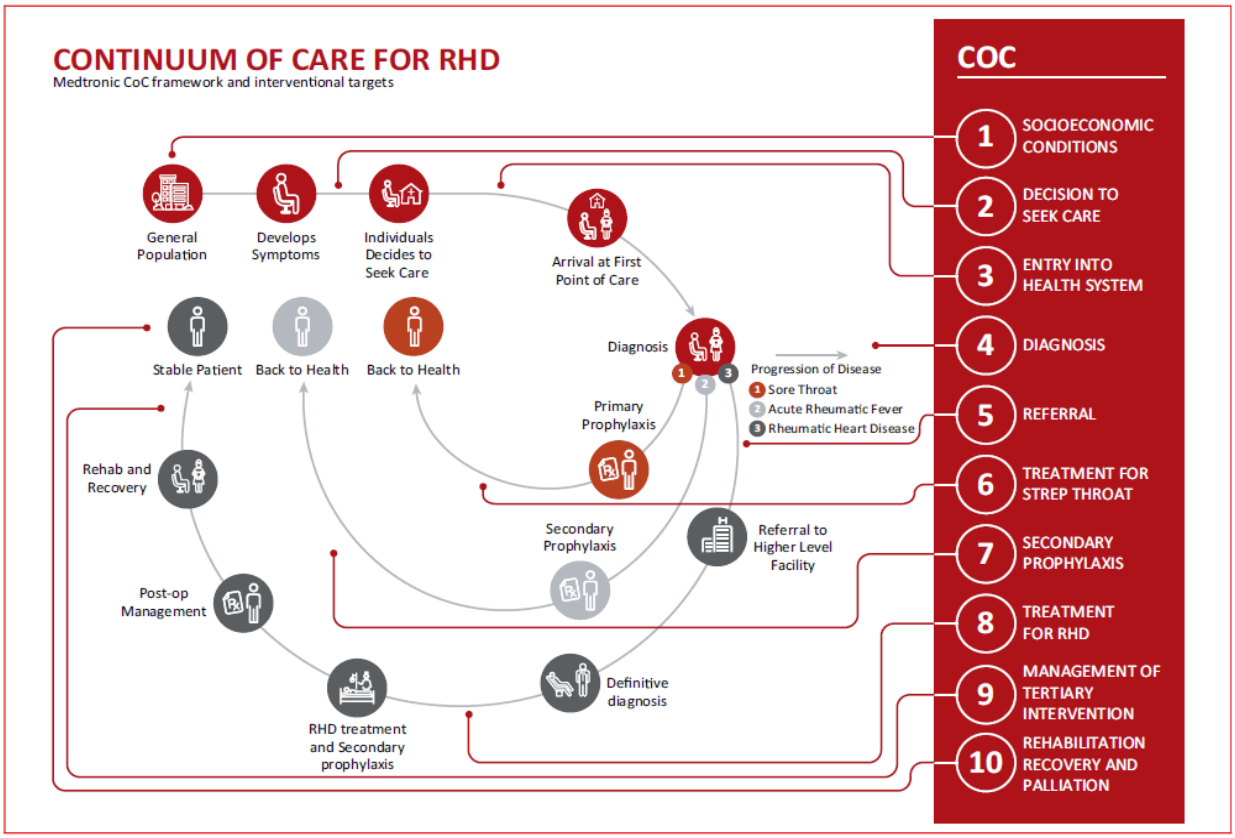


Figure 6.1 Continuum of Care for RHD *(Adapted from WHF 2017)*

As a patient-centered framework, the CoC facilitates an understanding of the care seeking process from the individual's or their caregiver's perspective, while permitting the systematic identification and categorization of the various needs and opportunities of patients, providers, communities and the wider health system to manage cases of GABHS sore throat, RF and RHD appropriately, and prevent the progression of disease. Hence there key points in the continuum of care for the health system to intervene to improve RHD care.

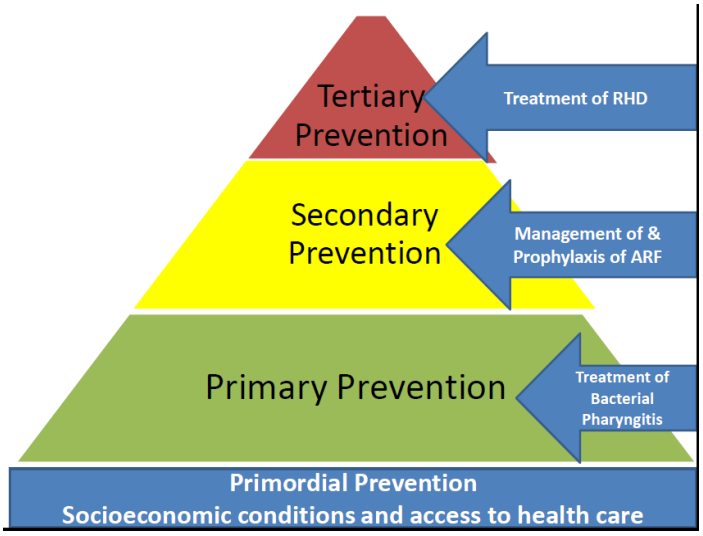
WHF developed quick tips for Comprehensive Prevention of RHD which recommends a comprehensive list of activities that need to be included in a RHD program. A framework has been developed to show this list of comprehensive activities (Figure 6.2).

	Advocacy		Research		
Tertiary Prevention	Medical management Of RF and RHD	Anticoagulation	Triage and preoperative planning	Postoperative planning	Provision of interventional services
Secondary Prevention	RF/RHD register	Supply of BPG and Other Antibiotics (erythromycin)	- Mitigate fear of anaphylaxis of BPG - Provision of secondary prophylaxis	Priority-based follow-up (clinical review)	Active case-finding (echocardiography screening)
Primary Prevention	Community education	Sore Throat Clinical diagnosis and treatment guidelines	Provision of primary prophylaxis	School-based interventions	Sore Throat register
Baseline and Health Systems	Government engagement	Disease notification	Human resources	Health worker training	Program evaluation
	Burden of disease data	RF/RHD Advisory Committee	Funding	Laboratory services	Integration with primary care & health systems
	Poverty Overcrowding Malnutrition Access to Health Care Hygiene				

Figure 6.2 Conceptual framework for comprehensive RF/RHD Prevention

(adapted from World Heart Federation 2013)

The framework deals with four levels interventions as summarized below.



Primordial prevention: address the underlying social determinants of health that exacerbate GAS exposure and RHD risk.

Primary prevention: treatment of strep throat to prevent the development of RF following GAS infection.

Secondary prevention: prevent development of new GAS infections following the first episode of acute RF to prevent subsequent RF recurrences and delay or prevent development of severe RHD.

Tertiary prevention: control RHD symptoms and extend the life of those living with RHD.

6.3 Barriers for RHD Prevention and Control Program

Even though cost effective interventions are available for the prevention and control of RHD, there are several barriers that hampered implementation of this program in Ethiopia and elsewhere.

1. **Health System-Related Barriers**
    - Lack of financial resources
    - Low priority given to RF/RHD
    - No development partner supporting RHD Program
    - Inadequate infrastructure
    - Human resources shortage
    - Health care workforce attrition
    - Poor capacity to detect RF and RHD (lack of echocardiography, lab tests, ECG)
    - Inadequate coordination and lack of team work
    - Absence of standardised clinical guidelines,
    - Shortages and stock outs of essential antibiotics, including quality BPG
    - Poor referral linkages
2. **Health Care Provider Related Barriers**
    - Inadequate training on Sore throat, Acute Rheumatic Fever and RHD diagnosis and management.
    - Poor understanding among health professionals about the purpose of treating sore throat and the link with RHD
    - Concerns about anaphylaxis, particularly among asymptomatic children, may make some health professionals reluctant to administer BPG and in some settings this has led to bans on Benzathine penicillin administration,
    - High work load on HCWs
    - Poor communication skills,
    - Low motivation of staff

3. Patient-related Barriers:

- Lack of access to health services
- Poverty
    - lack of transportation
    - Cost of medicines and investigations
- Lack of family support
- Harmful cultural and traditional practices
- Sore throat may not be considered an illness that warrants medical care, or its link with RF/RHD may be poorly understood, consequently patients/parents do not seek care

- Poor adherence may also arise due to pain from regular injections,
- Fear of allergy of drugs
- Severity of RHD limiting self care
- Stigma about heart disease
- Depression and substance use

The health system needs to identify these key barriers and solve them in collaboration with different stakeholders for effective implementation of the program.

6.4 National RHD Prevention and Control Program

6.4.1 Goals of RHD Program in Ethiopia:

- To reduce the occurrence and severity of Group A Beta Hemolytic Streptococcal (GABHS) infection and its suppurative and non-suppurative complications.
- To reduce morbidity, disability and mortality caused by RF/RHD and its complications in Ethiopia.

6.4.2 Specific Objectives:

- To increase the **awareness about RF/RHD to physicians, health officers, nurses, health extension workers, teachers and the public.**
- **Training** of health personnel including physicians, health officers, nurses, health extension workers on sore throat and relation with RHD, secondary prophylaxis, and management of RHD**.**
- Establishing a national **registry for RHD.**
- Establishing **screening programs for RHD for school children**
- Establishing strong **advocacy through mass media, health facilities, schools and health extension program.**
- Consolidation of **primary and secondary prevention through awareness and training.**
- Ensuring the continuous supply of good **quality benzathine penicillin.**
- To foster **research that can have an impact on RHD control**
- To strengthen partnership with national, regional and international **organizations concerned with RHD control.**

6.4.3 Recommended elements of a national RHD program

(based on the World Health Organization)

- 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

6.4.4 Strategies for RHD Control in Ethiopia:

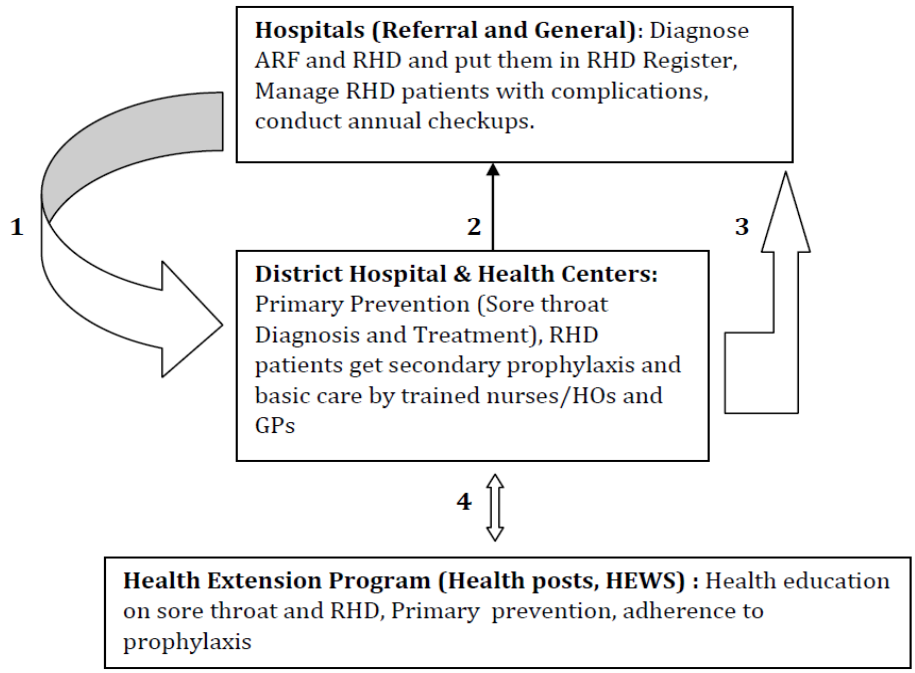


Figure 6.3 Pictorial presentation of patient referral and flow of care

6.4.5 Key Activities of RHD Program

1. Training and Support for Health Workers

Standardize guidelines for diagnosis and management of ARF and RHD

- Revised Jones Criteria for diagnosis of ARF
- Standardise dosing and delivery of secondary prophylaxis

Train health workers

- Curriculum development
- Training programmes for students and trained staff
- Updates for staff in rural and remote areas

Communicate

- Referral of new cases to Health center for ongoing care

- Update staff about on local ARF/ RHD issues
- Report on RHD in the community

2. Sore Throat Diagnosis

3. Identify & Register cases

Collect information on known cases of ARF & RHD

- Benzathine Penicillin injection books and clinic records
- Echocardiogram reports and cardiac surgery lists
- Hospital admission & death reports

Identify new cases

- Health centres or hospital when individuals present with ARF or RHD
- School health (screening) programmes
- Antenatal Clinics

Maintain a paper or computer register of all people with confirmed or suspected ARF & RHD

4. Refer patients to Hospital if there is

- Fever with arthritis or severe arthralgia (which prevented the patient from walking).
- Palpitations.
- Shortness of breath, unable to play with peers, or fainting
- Any Heart murmur
- Chorea (abnormal movement of extremities)

5. Screening for RHD

Considerations for screening

- Who to be screened *(e.g. school children are easier / but RHD may be more common in adults)*
- Methods of diagnosis available *(e.g. auscultation & clinical assessment, echocardiogram)*
- Availability of trained staff

- Processes for reporting RHD cases to the RHD programme
- Health resources available for long-term management of more RHD cases.

6. Optimize Secondary Prophylaxis

Establish secondary prophylaxis delivery

- Identify people who need secondary prophylaxis
- Identify health facilities where individuals receive secondary prophylaxis
- Supply of high quality Benzathine penicillin G

Improve secondary prophylaxis – identify specific barriers to treatment

- Identify people who do not receive adequate Benzathine penicillin injections (>80% injections)
- Establish recall and reminder systems
- Support communication between health facilities
    - *Refer new cases to peripheral health facilities for ongoing management*
    - *Identify people who move between health centers for treatment*

7. Community Support

Educate and Inform

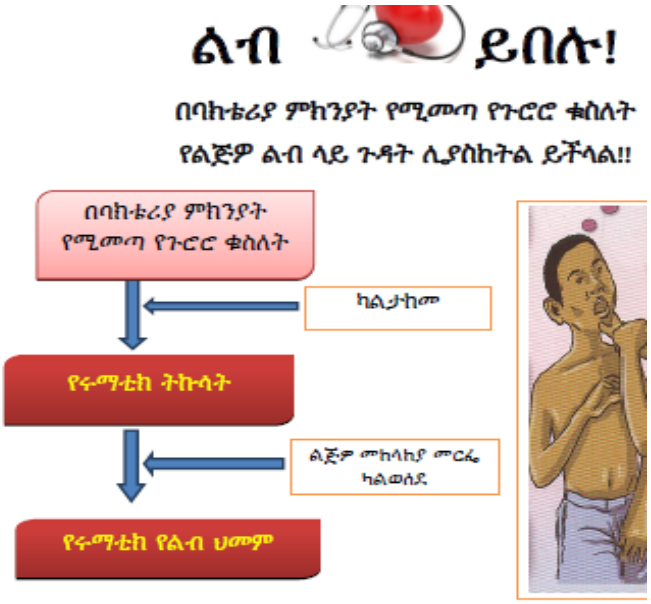
- Ensuring patients, parents, schoolteachers, care givers and communities are aware of the risks posed by untreated sore throat, RF and RHD through campaigns
- Community education materials *(posters & brochures)*

Manage individuals with ARF & RHD

- Promote ongoing medical care / echocardiogram / pregnancy counseling / dental care

Prioritize treatment for severe cases

- Cardiac assessment
- Surgery and support



6.4.6 Key Achievements of National RHD Program to date

- National Major NCDs Guideline developed
- National RHD TWG Established
- RHD Implementation action plan developed
- RHD Training material developed
- RHD patient level formats (Intake form, follow up form, RHD Patient passport) developed
- National RHD Register developed
- Client education materials developed

6.4.7 Things to do to strengthen the RHD Program

- Establish RHD Task force/TWG at RHB Level
- Assign RHD Focal Persons at Hospitals
- Select trainees for hospital based training
- Hospital based training for hospital and HC Staff
- Supply Recording and reporting tools
- Supply/quantification of Essential Medicines
- Sensitization and orientation of staff on strep sore throat, RF, Safe BPG injection.
- Orient staff on safe injection practices
- Referral linkage
- Mentoring
- Conduct Catchment area meetings
- Reporting
- Orientation and support of health extension workers
- Support activities in school health program

Unit 6 Summary

An RHD Programme should have:

- Government commitment
- A manageable RHD Register
- Well-trained, dedicated staff at all levels
- Systems to identify known cases and refer of new cases
- A priority system for severe cases
- Secondary Prophylaxis monitoring and improvement
- Ongoing support for health staff and the community.

Unit 7: Monitoring and Evaluation of RHD Program

Learning Objectives

At the end this module the participants will be able to:-

- Describe what is monitoring and evaluation
- Explain data management steps
- Describe the data capturing and aggregation tools for RHD Prevention and Control Services
- Introduce the draft tools and their use
- Discuss indicators for RHD Control program

7.1 Introduction

Key terms in Monitoring &Evaluation

Monitoring: is a routine tracking of progress toward the performance standards that were set.
Recording: is the practice of capturing data on patients' management over time and across clinical sites and writing information either directly on paper forms and/or entering into a computer.

Reporting: The routine tracking (monitoring) of priority program management information and its intended aggregated patient outcome data (evaluation) at the facility, Woreda, zone, region & national level

Evaluation: is the periodic assessment of programme impact and value.

Steps in data Management

- Data source- identify a data source
- Data collection- verify and collect data
- Data collation- add up the data
- Data Analysis –analyze the data based on preplanned indicators and targets
- Data interpretation- interprete the data
- Data presentation- present the data
- Data reporting- report or disseminate the data
- Data use –use the data for program improvement.

7.2 Recording and Reporting Tools in RHD Program in Ethiopia

A. Data capturing tools

- Patient Chart
- RHD Intake form
- RHD Follow up form
- Client-held RHD Passport
- Benzathine Penicillin injection Logbook

Data Aggregation tools

- RHD TREATMENT Register

Other

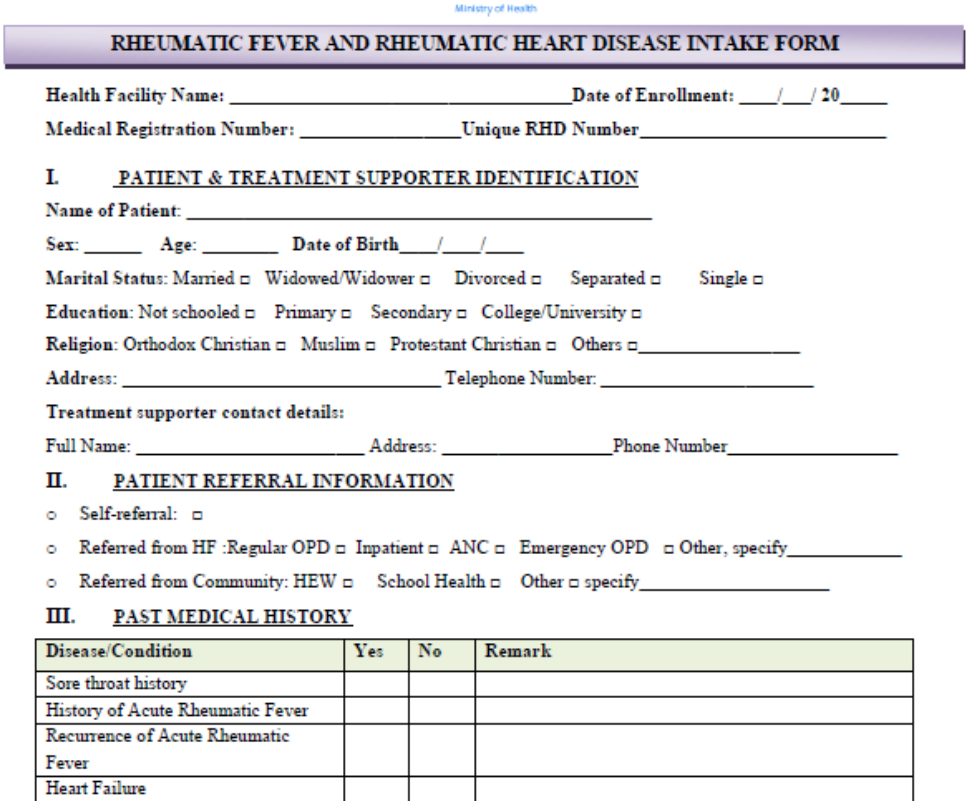
- Appointment logbook
- RHD reporting tools

7.3 Patient Level Recording Tools

A. RHD Intake Form

This is a 2 page format which helps in capturing all relevant information of the patient at **enrollment** to care.

It contains demographic data; history of current and past illnesses and treatments; current symptoms; anthropometric measures and vital signs; key physical findings and laboratory and investigations; final diagnosis and treatment plan.



Ministry of Health

RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE INTAKE FORM

Health Facility Name: _____**Date of Enrollment:** ____/___/ 20_____

Medical Registration Number: _____**Unique RHD Number**_____

I. PATIENT & TREATMENT SUPPORTER IDENTIFICATION

Name of Patient: _____

Sex: _____ **Age:** _____ **Date of Birth**____/____/____

Marital Status: Married ◻ Widowed/Widower ◻ Divorced ◻ Separated ◻ Single ◻

Education: Not schooled ◻ Primary ◻ Secondary ◻ College/University ◻

Religion: Orthodox Christian ◻ Muslim ◻ Protestant Christian ◻ Others ◻_____

Address: _____ Telephone Number: _____

Treatment supporter contact details:

Full Name: _____ Address: _____Phone Number_____

II. PATIENT REFERRAL INFORMATION

- Self-referral: ◻
- Referred from HF :Regular OPD ◻ Inpatient ◻ ANC ◻ Emergency OPD ◻ Other, specify_____
- Referred from Community: HEW ◻ School Health ◻ Other ◻ specify_____

III. PAST MEDICAL HISTORY

Disease/Condition	Yes	No	Remark
Sore throat history			
History of Acute Rheumatic Fever			
Recurrence of Acute Rheumatic Fever			
Heart Failure			

B. RHD Follow UP Form

This is also a two page format which helps the clinician in the follow up of patients in the routine care of RHD patients.

The first page captures the follow up parameters while the second page contains information on how to fill the format.

Federal Ministry of Health, Ethiopia

Rheumatic Fever/ Rheumatic Heart Disease Follow Up Form

Name of the Patient: _____ Age: _____ Sex_____ M.R.N._____ Unique RHD No. _____

Address: _____ Telephone No. _____ Date of Enrollment _____/_____/_____

Current Disease Status: Acute Rheumatic Fever_____ Mild RHD_____ Moderate RHD____ Severe RHD_____ NYHA_____

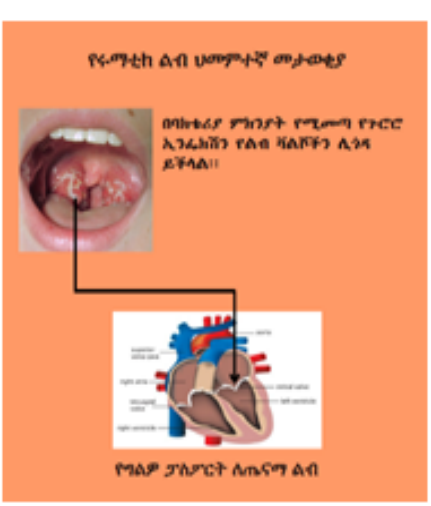
Status at Registration: New Case_____ Known case_____

Visit Type *Scheduled Unscheduled*	Date of Visit (__/__/__)	History	Anthropometry and Vital signs				Physical Examination				Investigations and Lab Tests					Treatment Plan and Adherence				Referral/ Consultations	Clinician's Name	Next Appointment __/__/__
		Current Symptoms	Weight (kg)	BMI (kg/m²)	BP (mmHg)	Pulse	Oral/ Dental	CVS	MSS	CNS	ECG	Echo	Hgb/ESR	Urinalysis	INR	BPG	Adherence to BPG	Warfarin	Other			
(a)	(b)	(c)	(d)	(e)	(f)	(g)	(h)	(i)	(j)	(k)	(l)	(m)	(n)	(o)	(p)	(q)	(r)	(s)	(t)	(u)	(v)	(w)

C. Client held RHD Patient Passport

This is a small booklet that contains patient and health facility information like name and address and unique RHD number of the client. Additionally it contains basic information on RHD.

It has a section for Benzathine penicillin injection which contains the date of injection, dose, batch number and who gave the injections and next appointment.



7.4 RHD Treatment Register

Do we need an RHD Register?

Yes. But why?

- Coordinate secondary prophylaxis and follow-up programmes
- Identify high risk individuals who require priority care
- Help identify others who may be at risk
- Provide information on the local rates of disease.

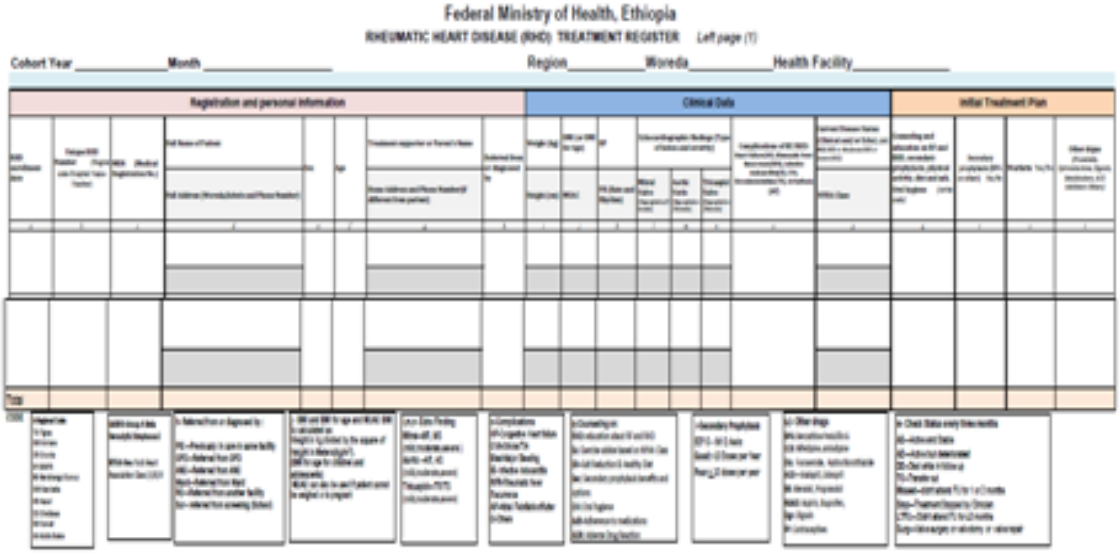
FMOH RHD Treatment Register

The national RHD Treatment Register has four parts:

- Cover page
- Page 1
- Page 2
- Back page

A. Page 1 of RHD Register has three sections

- **Header**
  - Cohort Year (write Month and year in Ethiopian Calendar, e.g Meskerem 2010)
- **Main Body** has three major headings:
  - **Registration and personal information:** Columns a-h
  - **Clinical Data:**Columns i-p
  - **Initial Treatment Plan:** Columns q-z:
- **Footer**: contains codes on how to fill the information in each column.



Registration and personal information contains the following information

- RHD enrollment date: write in Eth Calendar
- Unique RHD Number (Region code-Hospital Name- Number) e.g. TG-Ayder-00001
- MRN (Medical Registration No.)
- Full Name of Patient and address
- Sex and age
- Treatment supporter or Parent's Name and phone
- Referred from or diagnosed by.

Clinical Data:

- Weight, Height, BMI, MUAC
- BP, Pulse Rate, rhythm
- Echocardiography findings if available (Type of lesion and severity)
- Complications of RF/RHD: Heart Failure(HF), Rheumatic Fever Recurrence(RFR), Infective endocarditis(IE), CVA, thromboembolism(TE), Arrhythmia (AF)
- Current Disease Status: (Clinical and/or Echo) *(ARF, Mild RHD or Moderate RHD or Severe RHD) and NYHA Class (or modified Ross class)*

Initial Treatment Plan

- Counseling and education on RF and RHD, secondary prophylaxis, physical activity, diet and salt, Oral hygiene *(write code)*
- Secondary prophylaxis (BPG or other) *Yes/No*
- Warfarin Yes/No
- Other drugs *(Furosemide, Spironolactone, Digoxin, Beta blockers, ACE Inhibitors Others)*

Footer: Codes

- b-Regional Code:
- h- Referred from or diagnosed by :
- j- BMI and BMI for age and MUAC
- l, m,n- Echo Finding
- o-Complications
- q-Counseling on:
- r-Secondary Prophylaxis
- s,t- Other drugs
- w- Check Status every three months.

B. RHD Treatment Register Page 2

It contains columns u to bf. This page is used for the follow up of patients with RHD for up to 27 months.

When finished can start a new register with the same baseline information entered linking with the old register.

It mainly helps to document:

- Secondary prophylaxis (BPG or other)
- Follow up Medications
- Every 3 months additionally assess Clinical status, INR
- Annually: assess Clinical status, INR and document any New Echo finding.

7.5 Rheumatic Fever and RHD Indicators

To properly monitor and evaluate Rheumatic fever and Rheumatic heart disease prevention and control program some key indicators need to be collected and interpreted. Some of the key indicators are listed below.

- # newly diagnosed with bacterial (GABHS) sore throat (based on clinical score)
- # newly diagnosed with confirmed or probable acute rheumatic fever
- # diagnosed with confirmed or probable recurrence of acute rheumatic fever
- # newly diagnosed with rheumatic heart disease based on echo and/or clinical
- # with RHD currently on secondary prophylaxis (BPG monthly injections)
- Proportion on secondary prophylaxis who took >80% of the injections in the past 1 year.
- Number on monthly BPG who developed reactions following the injections (mild, anaphylactic, vasovagal reactions)
- Number of RHD patients who developed major complications (Infective endocarditis, bleeding, heart failure requiring admission, stroke, pregnancy)
- Number with RHD on medical treatment for heart failure
- Number with RHD who died and reasons for death
- Number with RHD who need surgical interventions (Severe RHD by echo or clinically)
- Number with RHD who underwent surgical interventions (Valve replacement or valve repair).
- Number with RHD on anticoagulation

Unit 7 Summary

- A RF/RHD Register is required to document burden of disease, to understand the need for advanced care and surgery, to introduce screening and secondary prophylaxis programs
- The information should be confidential and secure
- The information on the register should
  - Help coordinate health care for individuals
  - Help describe the level of disease in the community
- All confirmed and suspected cases of ARF and RHD should be notified.

ANNEXES

Annex 1: Check list for mouth, throat and dental examination

RESPONSE OPTION	YES	NO	PARTIAL	ASSESSMENT
wash your hands with • alcohol or soap &water for 15 sec • before touching the patient • after finishing				
Introduce yourself to patient /care taker and describe what you are going to do				
position the patient comfortably				
vital sign: temperature • clean thermometer • keep in axilla • inform mother or care taker or pt self-hold it tightly • wait for 3 min • read the temperature • clean the thermometer				
Examination of throat • Ask the patient to open the mouth wide • Examine back of tongue and tonsils (press down on the tongue with a tongue depressor) and look for o erythema of tonsil o swelling o follicle exudates o Examine teeth and gums				
palpation of anterior cervical lymph nodes				

Not observed *: steps ,task or skill not performed by participant during evaluation by trainer*
Learners *: study this tool to learn about and practice the correct steps needed to provide the clinical skill how to examine tonsils and paplapate cervical lymphnodes. Ask your colleagues to use the check list while you are parcticing among yourself and later with client .Specific feed back using this stool to guide the observation has to be given by the observer.*
Place in the box √ if taslk is performed satisfactorly , an ***X*** *if it is not performed satisfactory, N/O if the activity is not observed.*
Satisfactory *: perform the steps or task according to the standard procedure or guidelines*
Unsatisfactory *: Unable to perform the steps or task accoeding to the standardprocedure or guidelines*
Not observed *: Steps or tasks not performed by the participant during evaluation by the trainer*

Annex 2: Check List for Cardiovascular System Examination

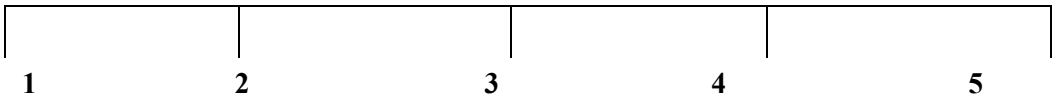
Activity	Yes	No	Assessment
Wash your hands			
Introduce yourself and describe what you are going to do			
Make ready your stethoscope and check itsfunctionality by listening to rubbing of your hands			
Warm your hand and your stethoscope by rubbing			
Position the patient comfortably with appropriate pillows under the head			
Make the room as quiet as possible			
Expose the patient anterior chest and the neck			
Precordium *Inspect the precordium for the presence of pulsation *Auscultate for the presence of abnormal heart sound(murmur) at the following positions: • Mitral : apex of heart • Tricuspid : Left fourth intecostal space next to sternum • Aortic : Right second intecostal space next to sternum			
General Appearance • Fast breathing • Restlessness • Unable to lie supine • Bluish discoloration of the lips or the tongue			
Vital signs: Check the following • Count radial pulse for a minute to know the rate. Also check regularity and volume of the pulse • Count respiratory rate • Take axillary temperature • Measure BP			
Neck Veins Inspect the neck for the presence of distended veins and multiple pulsations.			
Hepatomegaly Palpate RUQ for presence of liver enlargement and/or tenderness			
Edema Inspect ankle /leg for edema and check for pitting.			

ANNEX 3: DAILY COURSE EVALUATION FORM

Date _____

1. How valuable were today's sessions for you, based on a scale of 1 to 5?

(1 = not valuable at all; 5 = extremely valuable) **circle one**



2. Identify 3 things from today that will be useful to you in your job.
    1. _____
    2. _____
    3. _____

4. Please provide any comments or recommendations regarding the content of today's training.
    - _____
    - _____
    - _____

5. Please feel free to write any additional comments here.

_____

_____

_____

_____

_____

_____

ANNEX 4: COURSE EVALUATION FORM

Please indicate your opinion of the course components using the following rate scale:

5–Strongly Agree 4–Agree 3–No Opinion 2–Disagree 1–Strongly Disagree

COURSE COMPONENT	RATING
1. The classroom sessions were adequate for learning and clinical demonstration skills.	
2. The learning activities were helpful.	
3. There was sufficient time scheduled for practicing the demonstrations	
4. I can now be able to define ARF/RHD	
5. I am now able to differentiate bacterial tonsilitis from viral tonsilitis	
6. I am able discuss pathogenesis of ARF and RHD	
7. I am now confident in treating tonsilopharyngitis	
8. I am confident in diagnosing Acute Rheumatic fever	
9. I am now confident in Management and treatment of ARF	
10. I am now confident in identifying patients with RHD	
11. I can confidently discuss manifestations and complications of RHD	
12. I am no confident in following patients with ARF and RHD	
13. I can confidently discuss Commodity and data management for ARF/RHD	

ADDITIONAL COMMENTS (use reverse side if needed)

1. What topics (if any) should be **added** (and why) to improve the course

2. What topics (if any) should be **deleted** (and why) to improve the course?